FIFTH EDITION

PHYSICAL MEDICINE AND REHABILITATION BOARD REVIEW

SARA J. CUCCURULLO, MD
Editor-in-Chief

LESLIE BAGAY, MD
Associate Editor

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Physical Medicine and Rehabilitation Board Review

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Fifth Edition

Edited by

Sara J. Cuccurullo, MD

Professor and Chairman
Residency Program Director
Department of Physical Medicine and Rehabilitation
Hackensack Meridian School of Medicine
Rutgers Robert Wood Johnson Medical School
Physician in Chief
HMH Rehabilitation Care Transformation Services
Medical Director and Vice President
HMH JFK Johnson Rehabilitation Institute
Edison, New Jersey

Associate Editor

Leslie Bagay, MD

Clinical Associate Professor
Associate Director, Residency Program
Department of Physical Medicine and Rehabilitation
Hackensack Meridian School of Medicine
Rutgers Robert Wood Johnson Medical School
HMH JFK Johnson Rehabilitation Institute
Edison, New Jersey

Assistant Editor

Joseph Lee, MD

Clinical Assistant Professor
Department of Physical Medicine and Rehabilitation
Donald and Barbara Zucker School of Medicine at Hofstra/Northwell
Hempstead, New York

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I dedicate this book to three of the most important people in my life who have passed on:

My wonderful father, Pasquale Cuccurullo; his love, support, and encouragement are deeply missed since he passed away from lung cancer in 2004.

My amazing mother, Connie, who has been my support system throughout my entire life; her love, wisdom and friendship are missed every day since she passed in November 2022.

Also, my dear friend, Kathy Wong, MD. The spirit, integrity, and grace she brought to her patients and the field of Physical Medicine and Rehabilitation are greatly missed since she died of breast cancer at the young age of 36.

This book is also dedicated to:

My husband Alec, my loving partner in life;

My four children Michelle, Alexander, Amanda, and Nicholas, who are the joys of my life;

My mentors and teachers, especially Dr. Thomas E. Strax and Dr. Carolyn McCagg, my inspiration in all aspects of medicine, both clinical and academic, and Dr. Ernest W. Johnson, my encouragement to take on a challenge;

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Burns *Alan W. Young, DO, Kristen A. Harris, MD, and Shirley Shen, MD*
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FOREWORD

In memoriam—Ernie Johnson, MD
(1924–2014)

Dr. Johnson was a pioneer and a giant in Physical Medicine and Rehabilitation (PM&R) who always exhibited enormous energy. His contributions to our specialty are countless and impossible to measure. Back in 2001, he stated that our specialty of PM&R was lacking a PM&R Board Review book, and to fill that void he encouraged me to publish the notes from the JFK Johnson PM&R Board Review course that I had been teaching back then for over 10 years.

Dr. Johnson was extremely encouraging and supportive throughout every edition of the PM&R textbook, and wrote the forewords for the first, second, and third editions. In November 2014, Dr. Johnson passed away, and is sincerely missed. Prior to his death, he wrote the following excerpt of his support and enthusiasm for the third edition. In honor of him and all that he has done for our field, I have decided to include it as the foreword for this fifth edition of the PM&R Board Review textbook.

> This elegant volume finally fulfills a critical void and will supply reasonable and current PM&R diagnostic and management facts for the prospective board candidate.
>
> It can be studied in a reasonable time without speed reading *and* it is up-to-date with valuable and relevant information. The PM&R Board Review textbook has over 1,000 pages of nuggets. In addition, many physiatrists are coming up for recertification—certainly a major need for a PM&R comprehensive study and the solution is Dr. Cuccurullo's convenient and relatively inexpensive volume!
>
> **My prediction—the *PM&R Board Review* textbook is the optimal solution for studying for the board exams and recertification!**
>
> Thank you, Dr. Sara Cuccurullo!
>
> *Ernest W. Johnson, MD*

PREFACE

Physical Medicine and Rehabilitation Board Review, Fifth Edition, will appeal to medical students, residents, and practicing physiatrists. The book concentrates on board-related concepts in the field of Rehabilitation Medicine. Residents will find the book essential in preparing for Part I and Part II of the PM&R Board Certification because it is one of the only books of its kind with major focus on board-related material giving a synopsis of up-to-date PM&R orthopedic, neurologic, and general medical information all in one place. Over 500 diagrams simplify material that is board pertinent. In this way, important concepts are clarified and reinforced through illustration. All of the major texts of this specialty have been referenced to give the board examinee the most timely and relevant information and recommended reading.

The fifth edition differs from previous editions with the expansion of all chapters, especially within the Traumatic Brain Injury, Rheumatology, and Pain Medicine chapters, and the Musculoskeletal Spine, Cardiac, and Pulmonary subsections. In addition, all relevant epidemiology, treatments, and medications have been updated by the authors throughout the book. This book is clearly different from most texts. It is written in outline form and is about one-third the size of most textbooks. The topics are divided into major subspecialty areas and are authored by physicians with special interests and clinical expertise in the respective subjects. Board pearls are highlighted with an open-book icon throughout the text. These pearls are aimed at stressing the clinical and board-eligible aspects of the topics. This format was used to assist with last-minute preparation for the board examination and was inspired by the *Mayo Clinic Internal Medicine Board Review*. The contents are modeled after the topic selection of the *American Academy of Physical Medicine and Rehabilitation (AAPM&R) Self-Assessment Examination for Residents (SAE-R) Content Outline* (which is used by residents nationwide to prepare for the Self-Assessment Examination [SAE]). This was done specifically to help all residents, postgraduate years 2, 3, and 4, in yearly preparation and carry over from the SAE preparation to board exam preparation.

Two key points need to be addressed prior to using this text. This book is not a comprehensive textbook of PM&R. All chapters are prepared under the assumption that readers will have studied at length one or more of the standard textbooks of PM&R before studying this review.

My hope is that this text is a valuable tool to all physicians preparing for both the written and oral board exams, and also in managing issues of patient care. Practicing physiatrists should also find this book helpful in preparation for longitudinal learning. Because this is one of the first textbooks designed specifically for PM&R board preparation, the authors welcome any ideas for improvement from any of the readers. We wish you all the best in your studies.

Sara J. Cuccurullo, MD

ACKNOWLEDGMENTS

I've had the pleasure of helping residents learn what they need to know for their Physical Medicine and Rehabilitation (PM&R) Boards at JFK Johnson Rehabilitation Institute for more than 25 years. Over these years, I have had many requests for my yearly revised notes from former residents and from residents outside our program. For this reason, I gathered together an expert group of knowledgeable physicians to put together a comprehensive *PM&R Board Review* text. After the first edition was published, it was realized that improvements would make this text better with additions, updates, and needed alterations to the existing text. The second edition was an improved version of *PM&R Board Review*. The third edition was further improved, updated, and expanded to include new, highly relevant board topics such as Pain Management, Ethics, Ultrasound, and Palliative Care. The fourth edition was further updated, improved, and expanded in areas that have become more board-relevant, including Ultrasound, Cancer, and Rheumatology, which were revamped and included PM&R-relevant updated treatments and diagnostic criteria. Color was also added to the fourth edition to add definition and organization to the text. The fifth edition is further improved upon and expanded, especially within the Traumatic Brain Injury, Rheumatology, and Pain Medicine chapters, and the Musculoskeletal Spine, Cardiac, and Pulmonary subsections. I want to thank all those individuals who reached out to me to point out edits and subject matter inclusion that would improve this fifth edition textbook.

PM&R Board Review, Fifth Edition, reflects the commitment of the authors and the faculty at Rutgers Robert Wood Johnson Medical School and Hackensack Meridian School of Medicine in the Department of PM&R based at JFK Johnson Rehabilitation Institute to produce a text that would be used as a guide containing selected topics considered important for physicians preparing for the certifying or the recertifying examination, or longitudinal learning, offered by the American Board of Physical Medicine and Rehabilitation (ABPMR). This text hopefully presents clear practical information for both residents studying for the boards of PM&R and for practicing physicians. This text should be of great value in not only preparing for the ABPMR board exam but also caring for patients.

Thanks for this textbook coming to print is given to Thomas Strax, MD. His encouragement and willingness to support this project from the start has been an inspiration in seeing this textbook come to realization. Special thanks have to be given to the administration of HMH JFK Johnson Rehabilitation Institute and HMH JFK University Medical Center for their encouragement and financial support, without which this book would not have been possible. Specifically, I would like to sincerely thank Anthony Cuzzola, J. Scott Gebhard, Michele Morrison, and Dr. Michael Kleiman, in addition to our HMH leadership Bob Garrett, Mark Sparta, Regina Foley, Ken Sable, Mark Stauder, David Kountz, MD, Ihor Sawczuk, MD, Jim Clarke, Dan Varga, Jose Lozano, and Michael McCauley. I would also like to thank Jeff Boscamp, MD, Dean of Hackensack Meridian School of Medicine, and Amy Murtha, MD, Dean of Rutgers Robert Wood Johnson Medical School, who support each and every academic endeavor put forth from our Department of PM&R.

I will also be eternally grateful to four of my former students, Joseph Lee, MD (my dedicated and tireless assistant editor), Edgardo Baerga, MD, Eric Freeman, DO, and Priscila Gonzalez, MD. It was their stamina and perseverance that enabled the first edition of this text to come to fruition. Their energy and enthusiasm were truly inspirational. I am grateful to all the authors for their completion of the manuscripts. I greatly acknowledge the support of Springer Publishing, specifically Beth Barry, Elizabeth Nieginski, Taylor Ball, John Zaphyr, and Kris Parrish.

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I would also like to thank Dr. Ernie W. Johnson, who has been very inspirational in any educational project I have undertaken. He is truly one of the giants in the field of PM&R. His support to write this *PM&R Board Review* textbook and his input prior to the publication of the first, second, and third editions are greatly appreciated. He is sincerely missed since he passed in 2014. I would like to acknowledge the enormous support and understanding I have received from my husband and four children during the formulation of this new edition. It is my sincere hope that *Physical Medicine and Rehabilitation Board Review*, Fifth Edition, will receive a warm reception. My coauthors and I look forward to receiving comments and suggestions from the readers.

Sara J. Cuccurullo, MD

CONTRIBUTORS

Jessica Ace, MD Pediatric Rehabilitation Attending Physician, Atlantic Health Children's Specialty Center, Morristown, New Jersey.

Michael Alexander, MD Professor, Departments of Pediatrics and Physical Medicine and Rehabilitation, Thomas Jefferson University, Philadelphia, Pennsylvania; Emeritus Medical Staff, Alfred I duPont Hospital for Children, Wilmington, Delaware.

Edgardo Baerga, MD Director, Stroke Rehabilitation Program, Encompass Health Rehabilitation Hospital of Tinton Falls, Tinton Falls, New Jersey.

Leslie Bagay, MD Clinical Associate Professor, Associate Director, Residency Program, Director of Medical Student Education, Department of Physical Medicine and Rehabilitation, Hackensack Meridian School of Medicine, Rutgers-Robert Wood Johnson Medical School, Medical Director, Cancer Rehabilitation Program, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

David P. Brown, DO Clinical Professor, Department of Physical Medicine and Rehabilitation, Hackensack Meridian School of Medicine, Rutgers Robert Wood Johnson Medical School; Director of Outpatient Services, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Lisa Casale, MD Assistant Professor, Department of Internal Medicine, Hackensack Meridian School of Medicine, Edison, New Jersey.

Jessie Chan, MD Chief Resident Physician, Department of Physical Medicine and Rehabilitation, Rutgers Robert Wood Johnson Medical School, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Jennifer Chui, MD Clinical Assistant Professor, Department of Physical Medicine and Rehabilitation, Rutgers Robert Wood Johnson Medical School, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Michael Chung, MD Past Chief Resident Physician, Department of Physical Medicine and Rehabilitation, Rutgers Robert Wood Johnson Medical School, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey. Currently an Interventional Pain Fellow, Emory University School of Medicine, Atlanta, Georgia.

Sara J. Cuccurullo, MD Clinical Professor and Chair, Residency Program Director, Department of Physical Medicine and Rehabilitation, Hackensack Meridian School of Medicine, Rutgers Robert Wood Johnson Medical School; Physical in Chief for the Rehabilitation Care Transformation Services, Hackensack Meridian Health; Medical Director and Vice President, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Laurent Delavaux, MD Clinical Assistant Professor, Pain Medicine Fellowship Director, Department of Physical Medicine and Rehabilitation, Hackensack Meridian School of Medicine; Rutgers Robert Wood Johnson Medical School, Attending Physician, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Anna Maria Dunn, MD Past Clinical Associate Professor, Department of Physical Medicine and Rehabilitation, Rutgers Robert Wood Johnson Medical School, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Elie Elovic, MD Einstein Physical Medicine and Rehabilitation at Moss, Jefferson Health, Philadelphia, Pennsylvania.

Steven V. Escaldi, DO Clinical Associate Professor, Department of Physical Medicine and Rehabilitation, Hackensack Meridian School of Medicine, Rutgers Robert Wood Johnson Medical School; Medical Director, Spasticity Management Program, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Talya Fleming, MD Clinical Associate Professor, Department of Physical Medicine and Rehabilitation, Hackensack Meridian School of Medicine, Rutgers Robert Wood Johnson Medical School, Director of Stroke Recovery, Aftercare and Long-COVID Program, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Eric D. Freeman, DO Interventional Pain Management, Chief Medical Officer, Redefine Healthcare-Orthopedic Pain and Spine Center, Edison, New Jersey.

Ted L. Freeman, DO Medical Director, Freeman Orthopedics and Pain Center, Brick, New Jersey.

Michael Galibov, DO Chief Resident Physician, Department of Physical Medicine and Rehabilitation, Rutgers Robert Wood Johnson Medical School, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Priscila Gonzalez, MD Mid Atlantic Rehabilitation Consultants, LLC, Encompass Rehabilitation Hospital of Tinton Falls, Tinton Falls, New Jersey.

Phillip Gordon, MD Past Chief Resident Physician, Department of Physical Medicine and Rehabilitation, Rutgers Robert Wood Johnson Medical School, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey. Currently a Spinal Cord Injury Medicine Fellow, Mount Sinai Hospital, New York, New York.

Christine Greiss, DO Clinical Associate Professor, Department of Physical Medicine and Rehabilitation, Hackensack Meridian School of Medicine, Rutgers Robert Wood Johnson Medical School; Medical Director of Concussion Program, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Kristen A. Harris, MD Clinical Assistant Professor, Department of Physical Medicine and Rehabilitation, Hackensack Meridian School of Medicine, Rutgers Robert Wood Johnson Medical School, Medical Director of Extended Recovery Unit, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Eric Heckelmann, MD Past Chief Resident Physician, Department of Physical Medicine and Rehabilitation, Rutgers Robert Wood Johnson Medical School, Attending Physician, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Barbara Hoffer, DO Physiatrist, Reading, Pennsylvania.

Bart K. Holland, PhD Professor Emeritus, Biostatistics, Rutgers New Jersey Medical School, Newark, New Jersey.

Beverly Hon, MD Clinical Assistant Professor, Department of Physical Medicine and Rehabilitation, Rutgers New Jersey Medical School, Newark, New Jersey; Staff Physician, Department of Spinal Cord Injury, VA NJ Health Care System, East Orange, New Jersey

Iqbal Jafri, MD Clinical Professor, Associate Program Director of Pain Medicine Fellowship Program, Department of Physical Medicine and Rehabilitation, Hackensack Meridian School of Medicine, Rutgers Robert Wood Johnson Medical School; Medical Director of Inpatient Cardiac Rehabilitation Program and Medical Director of Interdisciplinary Chronic Pain Program, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Faraz Jamal, DO Chief Resident Physician, Department of Physical Medicine and Rehabilitation, Rutgers Robert Wood Johnson Medical School, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Ernest W. Johnson, MD‡ Professor Emeritus, Department of Physical Medicine and Rehabilitation, College of Medicine, The Ohio State University, Columbus, Ohio.

Jaclyn Joki, MD Clinical Assistant Professor, Department of Physical Medicine and Rehabilitation, Hackensack Meridian School of Medicine; Rutgers Robert Wood Johnson Medical School; Director, PM&R Consult Service at Robert Wood Johnson University Hospital, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Stacey Jou, MD Past Chief Resident Physician, Department of Physical Medicine and Rehabilitation, Rutgers Robert Wood Johnson Medical School, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey. Currently a Brain Injury Medicine Fellow, Moss Rehab, Einstein Medical Center Elkins Park, Elkins Park, Pennsylvania.

Steven Kirshblum, MD Professor and Chair, Department of Physical Medicine and Rehabilitation, Rutgers New Jersey Medical School, Newark, New Jersey; Chief Medical Officer, Kessler Institute for Rehabilitation; Chief Medical Officer, Kessler Foundation, West Orange, New Jersey.

Joseph Lee, MD Assistant Professor, Department of Orthopedics, Department of Rehabilitation and Human Performance, Mount Sinai Hospital, New York, New York.

Christopher Leonardi, MD Past Rutgers Robert Wood Johnson Medical School medical student. Currently a Resident Physician, Department of Physical Medicine and Rehabilitation, University of Utah, Salt Lake City, Utah.

Jing Liang, MD Illinois Bone and Join Institute, Barrington, Illinois.

Lei Lin, MD, PhD Clinical Associate Professor, Co-Director of Quality Improvement, Department of Physical Medicine and Rehabilitation, Hackensack Meridian School of Medicine, Rutgers Robert Wood Johnson Medical School, Attending Physician, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Lisa Luciano, DO Past Clinical Associate Professor, Department of Physical Medicine and Rehabilitation, Rutgers Robert Wood Johnson Medical School, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Ofure Luke, MD Clinical Assistant Professor, Department of Physical Medicine and Rehabilitation, Hackensack Meridian School of Medicine, Rutgers Robert Wood Johnson Medical School; Attending Physician, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Ian B. Maitin, MD, MBA Professor and Vice Chair, Physical Medicine and Rehabilitation, Neuromusculoskeletal Institute, Rowan Virtua School of Medicine; PM&R Residency Program Director, Inspira Health System, Stratford, New Jersey.

Seema Malkana, DO Assistant Professor, Division of Rheumatology and Connective Tissue Research, Rutgers Robert Wood Johnson Medical School; Associate Program Director, Rheumatology Fellowship, Robert Wood Johnson University Hospital/RWJ Barnabas Health, New Brunswick, New Jersey.

Steven Markos, MD Assistant Professor, Department of Physical Medicine and Rehabilitation, Hackensack Meridian School of Medicine; Rutgers Robert Wood Johnson Medical School, Attending Physician, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

‡Deceased

Nicholas G. Melillo, MD Associate Clinical Professor and Co-Director, Critical Care Fellowship, Seton Hall School of Graduate Medical Education, South Orange, New Jersey; Attending Physician, Internal Medicine, Pulmonary Disease and Critical Care Medicine, HMH JFK Medical Center, Edison, New Jersey.

Matthew Moore, DO Assistant Professor, Department of Physical Medicine and Rehabilitation, Hackensack Meridian School of Medicine, Rutgers Robert Wood Johnson Medical School, Attending Physician, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Jeremiah Nieves, MD Clinical Assistant Professor, Associate Director of Spinal Cord Injury Medicine Fellowship, Rutgers New Jersey Medical School Department of Physical Medicine and Rehabilitation, Kessler Institute for Rehabilitation, West Orange, New Jersey.

Thomas R. Nucatola, MD Attending Rheumatologist, Institute for Rheumatic and Autoimmune Diseases-South, Clark, New Jersey.

Sagar Parikh, MD Clinical Assistant Professor, Pain Medicine Fellowship Associate Program Director, Department of Physical Medicine and Rehabilitation, Hackensack Meridian School of Medicine, Rutgers Robert Wood Johnson Medical School; Attending Physician, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Hayk Petrosyan, PhD Associate Professor, Department of Physical Medicine and Rehabilitation, Hackensack Meridian School of Medicine; Research Scientist, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Jonathan Quevedo, MD Past Clinical Associate Professor, Department of Physical Medicine and Rehabilitation, Rutgers Robert Wood Johnson Medical School, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

David S. Rosenblum, MD Medical Director, Milne Institute for Healthcare Innovation, Gaylord Hospital; Medical Director, Gaylord Hospital Spinal Cord Injury Program, Wallingford, Connecticut.

Roger Rossi, DO Clinical Professor, Department of Physical Medicine and Rehabilitation, Hackensack Meridian School of Medicine, Rutgers Robert Wood Johnson School of Medicine; Medical Director Acute Rehabilitation Program, HMH/Saint Joseph's Health; Director, Parkinson's Disease and Movement Disorders Program, HMH/Saint Joseph's Health, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Casey Schoenlank, MD Clinical Assistant Professor, Department of Physical Medicine and Rehabilitation, Hackensack Meridian School of Medicine; Director of Rehab Medicine Consult Services, HMH OUMC Johnson Rehabilitation Institute, Brick, New Jersey.

Richard M. Schuman, MD Assistant Clinical Professor of Medicine, Rutgers Robert Wood Johnson Medical School; Medical Director of Oncology, HMH JFK Medical Center, Edison, New Jersey.

Mary T. Shea, MA, OTR, ATP Clinical Specialist, Kessler Institute for Rehabilitation, West Orange, New Jersey; Adjunct Professor, New York University, New York, New York.

Shirley Shen, MD Chief Resident Physician, Department of Physical Medicine and Rehabilitation, Rutgers Robert Wood Johnson Medical School, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Alexander Shustorovich, DO Associate Program Director, Pain Medicine Fellowship; Clinical Assistant Professor, Department of Physical Medicine and Rehabilitation, Hackensack Meridian School of Medicine; Rutgers Robert Wood Johnson Medical School, Attending Physician, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Michelle Sirak, MD Chief, Pediatric Physical Medicine and Rehabilitation, Joseph M. Sanzari Children's Hospital, Hackensack Meridian Health, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Thomas E. Strax, MD Professor Emeritus, Department of Physical Medicine and Rehabilitation, Rutgers Robert Wood Johnson Medical School, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Krishna J. Urs, MD Clinical Assistant Professor, Co-Director of Quality Improvement, Department of Physical Medicine and Rehabilitation, Hackensack Meridian School of Medicine, Rutgers Robert Wood Johnson Medical School; Medical Director of JFK Medical Center Consult Services, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Heikki Uustal, MD Clinical Associate Professor, Department of Physical Medicine and Rehabilitation, Hackensack Meridian School of Medicine, Rutgers Robert Wood Johnson Medical School; Director of Prosthetics and Orthotics Team and Lab, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Craig Van Dien, MD Clinical Assistant Professor, Department of Physical Medicine and Rehabilitation, Hackensack Meridian School of Medicine; Rutgers Robert Wood Johnson Medical School; Medical Director for Center for Sports and Spine Medicine, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Lyn Weiss, MD Chair, Physical Medicine and Rehabilitation, NYU Langone Hospital–Long Island; Professor of Rehabilitation Medicine, NYU Grossman Long Island School of Medicine, Mineola, New York.

Alan W. Young, DO Chief, Complementary and Integrative Medicine Clinic, Interdisciplinary Pain Management Service, Department of Rehabilitation Medicine, Brooke Army Medical Center; Consultant, Rehabilitation Services, United States Army Institute of Surgical Research (Burn Unit), Fort Sam Houston, Texas.

James Yun, MD Chief Resident Physician, Department of Physical Medicine and Rehabilitation, Rutgers Robert Wood Johnson Medical School, HMH JFK Johnson Rehabilitation Institute, Edison, New Jersey.

Yi (Dan) Zhou, MD Past Chief Resident Physician, Department of Physical Medicine and Rehabilitation, Rutgers Robert Wood Johnson Medical School, HMH Johnson Rehabilitation Institute, Edison, New Jersey. Currently a Brain Injury Medicine Fellow, Spaulding Rehabilitation Hospital, Harvard Medical School, Boston, Massachusetts.

Richard D. Zorowitz, MD Chief Medical Informatics Officer, MedStar National Rehabilitation Network; Professor of Clinical Rehabilitation Medicine, Georgetown University School of Medicine, Washington, DC.

INTRODUCTION: BOARD CERTIFICATION

The discussion in this section is aimed primarily at candidates preparing for the American Board of Physical Medicine and Rehabilitation (ABPMR) certification examination or Maintenance of Certification© (MOC) examinations in Physical Medicine and Rehabilitation (PM&R). The following information was collected from and calculated by the ABPMR and is available on the ABPMR website at www.abpmr.org.

THE PURPOSE OF CERTIFICATION

The intent of the certification process as defined by Member Boards of the American Board of Medical Specialties (ABMS) is to provide assurance to the public that a certified medical specialist has successfully completed an accredited residency training program and an evaluation, including an examination process, designed to assess the knowledge, experience, and skills requisite to the provision of high-quality patient care in that specialty. Diplomates of the ABPMR possess particular qualifications in this specialty.

THE EXAMINATION

As part of the requirements for certification by the ABPMR, candidates must demonstrate satisfactory performance in an examination conducted by the ABPMR covering the field of PM&R. The initial examination for certification is given in two parts, Part I (computer-based) and Part II (oral).

Parts I and II of the Board examination are given once a year at times and places as the Board designates. Part I of the examination is administered simultaneously in August at Pearson Professional centers nationwide, while Part II is administered in the spring virtually via a video conferencing software.

EXAMINATION ADMISSIBILITY REQUIREMENTS

PART I ADMISSIBILITY REQUIREMENTS

The application and related forms are available on the physician homepage of the ABPMR website. The completed application must include a copy of the medical degree diploma or certificate as well as the postgraduate year 1 (PGY-1) certificate, if applicable.

In order to have the application considered for examination, the applicant must be scheduled to complete the graduate medical education requirements on or before August 31 immediately following the scheduled examination date for which they have applied. Satisfactory completion of the educational and training requirements in force at the beginning of the resident's training in an accredited program will be considered acceptable for application for admissibility to the certification examinations. Candidates who are engaged in the Clinical Research Pathway or who are pursuing Dual Specialty Certification should refer to the ABPMR website for more details.

Final admissibility is contingent upon receipt of the final-year evaluation by the program director, due August 1 in the examination year. In the final-year evaluation, the program director must affirm that the physician has satisfactorily completed PM&R residency training and has demonstrated sufficient competence to enter practice without direct supervision. The residency program director must recommend the physician for admissibility to the Part I examination. If a resident is placed on probationary status during the final year of the residency program, this status must be rescinded by the program director before July 1 in order for the resident to be admissible.

PART II ADMISSIBILITY REQUIREMENTS

Part II of the ABPMR certification examination is an oral examination. To be admissible for the examination, applicants must have passed Part I prior to applying for Part II. The application and related forms for Part II are available on the ABPMR website.

The applicant is required to submit a copy of a current, valid, and unrestricted license (including expiration date) to practice medicine in at least one jurisdiction in the United States, its territories, or Canada. Evidence of unrestricted licensure in the state or states where the physician practices will be required prior to issuance of the certificate.

REAPPLICATION

Physicians who have initially applied for and failed or did not take either Part I or Part II can apply for admissibility for reexamination or examination during any subsequent examination administration during the board eligibility period. The same requirements will be in effect for reapplication as for initial admissibility.

In accordance with ABMS recommendations, the board eligibility period was shortened to tighten the connection between training and certification. For physicians who completed training prior to January 1, 2012, the initial certification process must be completed by December 31, 2019. Physicians completing training on or after January 1, 2012, have seven calendar years after completion of residency to complete the initial certification process and become board-certified.

After the period of board eligibility has expired, candidates who have not successfully completed the initial board certification process can no longer identify themselves as board-eligible. Board eligibility status may be extended in certain circumstances; see the ABPMR website for further details.

THE EXAMINATION: PART I

Part I is a computer-based examination consisting of 325 multiple-choice questions divided into two 3-hour sections. There is a 60-minute break between sessions. An on-screen tutorial is available at the beginning of the first session, allowing the examinee to become familiar with both the computer and the format of the examination. The examination questions are designed to test the candidate's knowledge of basic sciences and clinical management as related to the PM&R field and will be in the form of objective testing.

Two forms of identification are required for examination: at least one government-issued identification with a signature and a recent photograph, and another form of identification that contains at least a signature. The name listed on both forms of identification must match the name on file with ABPMR. No notes, textbooks, other reference materials, scratch paper, or electronic devices may be taken into the examination room. Please refer to the ABPMR website for more detailed information about how to prepare for the Part I computer-based examination.

Part I of the certification exam outline consists of five content domains, and all test questions are classified into each of these domains.

PART I EXAMINATION OUTLINE

A. Foundational Rehabilitation Concepts (Across All Patient Ages) 12%

A.1. Diagnostic Procedures 2%
- **A.1.a Cardiovascular and pulmonary assessment**
- **A.1.b. Gait analysis**
- **A.1.c. Laboratory studies**
- **A.1.d. Medical imaging**
- **A.1.e. Musculoskeletal ultrasound**
- **A.1.f. Neuropsychological evaluation**
- **A.1.g. Urodynamics**

A.2. Treatments 3%
- **A.2.a. Electrical stimulation**
- **A.2.b. Integrative medicine**
- **A.2.c. Modalities**
- **A.2.d. Pharmacotherapy**
- **A.2.e. Procedural interventions**
- **A.2.f. Regenerative medicine**
- **A.2.g. Therapeutic exercise**

A.3. Adaptive or Assistive Technology 3%
- **A.3.a. Augmentative communication**
- **A.3.b. Assistive aids for mobility and activities of daily living**
- **A.3.c. Orthoses and braces**
- **A.3.d. Shoes**
- **A.3.e. Wheelchairs and seating**

A.4. Applied Science 4%
- **A.4.a. Anatomy and physiology**
- **A.4.b. Epidemiology and risk assessment**
- **A.4.c. Growth, development, and aging**
- **A.4.d. Kinesiology, biomechanics, and physical examination**
- **A.4.e. Mental and behavioral health**
- **A.4.f. Nutrition**
- **A.4.g. Pharmacology**
- **A.4.h. Research and statistics**

B. Electrodiagnostic Medicine (Across All Patient Ages) 10%

B.1. Electromyography
B.2. H reflex and F wave
B.3. Instrumentation
B.4. Nerve conduction
B.5. Neuromuscular transmission

C. Neurological Rehabilitation (Across All Patient Ages) 30%

C.1. Conditions 12%
- C.1.a. Brain injury
- C.1.b. Functional neurologic disorder (including conversion disorder)
- C.1.c. Motor neuron disorders
- C.1.d. Movement disorders (e.g., parkinsonism)
- C.1.e. Multiple sclerosis
- C.1.f. Myopathy, myositis, and dystrophy
- C.1.g. Neuromuscular junction disorders
- C.1.h. Neuropathy (e.g., mononeuropathy, polyneuropathy, plexopathy)
- C.1.i. Spinal cord injury
- C.1.j. Stroke

C.2. Dystonia and Spasticity 5%
- C.2.a. Chemodenervation
- C.2.b. Evaluation
- C.2.c. Intrathecal pumps
- C.2.d. Nonpharmacological interventions
- C.2.e. Oral medications
- C.2.f. Surgery

C.3. Rehabilitation Management Conditions 13%
- C.3.a. Agitation
- C.3.b. Cognitive disorders
- C.3.c. Contracture
- C.3.d. Dysautonomia
- C.3.e. Dysphagia
- C.3.f. Heterotopic ossification
- C.3.g. Hydrocephalus
- C.3.h. Neurogenic bladder and bowel
- C.3.i. Neuropathic pain
- C.3.j. Paralysis/paresis
- C.3.k. Posture, balance, and gait disorders
- C.3.l. Pressure injuries
- C.3.m. Reintegration
- C.3.n. Seizures
- C.3.o. Sleep disorders
- C.3.p. Speech and language disorders
- C.3.q. Visual dysfunction

D. Musculoskeletal Rehabilitation (Across All Patient Ages) 33%

D.1. Musculoskeletal and Occupational Medicine 20%
- D.1.a. Arthritides: inflammatory, noninflammatory, infectious
- D.1.b. Bone health
- D.1.c. Complex regional pain syndrome
- D.1.d. Fibromyalgia
- D.1.e. Fractures
- D.1.f. Muscle, ligament, and tendon disorders
- D.1.g. Myofascial pain
- D.1.h. Overuse syndromes
- D.1.i. Spinal disorders
- D.1.j. Thoracic outlet syndrome

D.2. Amputation 5%
- D.2.a. Upper extremity
- D.2.b. Lower extremity
- D.2.c. Prostheses

D.3. Rehabilitation Management Conditions 8%
- **D.3.a. Abnormal gait**
- **D.3.b. Contracture**
- **D.3.c. Deconditioning: atrophy and immobilization**
- **D.3.d. Pain**
- **D.3.e. Reintegration**

E. Medical Rehabilitation (Across All Patient Ages) 15%

E.1. Conditions 5%
- **E.1.a. Burns**
- **E.1.b. Cancer**
- **E.1.c. Cardiovascular disorders**
- **E.1.d. Endocrine and metabolic disorders**
- **E.1.e. Postinfectious disease**
- **E.1.f. Pulmonary disorders**
- **E.1.g. Transplant**

E.2. Rehabilitation Management Conditions 10%
- **E.2.a. Contracture**
- **E.2.b. Deconditioning**
- **E.2.c. Delirium**
- **E.2.d. Dysphagia**
- **E.2.e. Edema**
- **E.2.f. Malnutrition**
- **E.2.g. Medical complications**
- **E.2.h. Reintegration**
- **E.2.i. Sleep disorders**
- **E.2.j. Wound care**

QUESTION FORMAT

The 1998 ABPMR booklet gave an idea of how the exam looks. These items are not from previous ABPMR exams, nor will they appear on future tests. They are given by ABPMR as a sample for your use. All items are of the "best single choice answer multiple-choice" type. In June 2015, the ABPMR released "Part I Practice Questions," which contains 100 practice questions and is available on the ABPMR website. Example questions are noted as follows:

1. Postacute recovery and community reintegration of the traumatically brain-injured patient are most often hampered by:
   A. Language impairment
   B. Memory impairment
   C. Physical impairment
   D. Financial disincentives
   E. Personality and behavioral impairment
2. Which best describes a feature of short-wave diathermy?
   A. It is used to heat the hip joint.
   B. It produces both direct and reflex blood flow increase.
   C. It is used around the thigh to improve circulation in an ischemic limb.
   D. The dose is regulated by measuring the flow of the high-frequency current through the patient.
   E. Commercially available machines operate at a frequency of 950 MHz.

3. The single most reliable clinical sign for the detection of inflammatory arthritis is:
   A. Local tenderness
   B. Painful, limited range of motion
   C. Synovial swelling
   D. Joint effusion
   E. Skin color change
4. Which condition is most likely a contraindication for intra-articular corticosteroid injection therapy?
   A. Crystal-induced synovitis
   B. Diabetes mellitus
   C. Peptic ulcer
   D. Bacteremia
   E. Osteoarthritis

Answers for the previously mentioned examples are as follows; 1. E, 2. B, 3. C, 4. D. Attempts have been made to avoid ambiguity and typographical or spelling errors, but occasionally they occur. They are not intended to "trip you up" or confuse you.

THE EXAMINATION: PART II

The oral examination consists of two 50-minute sessions and involves an interactive process between the candidate and two examiners. One 10-minute break divides the two sections of the oral examination. Each 50-minute session consists of four patient vignettes.

During the Part II examination, each examiner will present a vignette comprising a clinical case scenario and subsequently ask questions about diagnostic procedures, therapeutic procedures, and patient management. Candidates will be expected to present, in a concise, orderly fashion, evidence of their proficiency in the management of various clinical conditions within the field of PM&R. Performance on each vignette is evaluated using performance criteria within five domains: data acquisition, problem-solving, patient management, systems-based practice, and interpersonal and communication skills. The examination content is classified according to Class 1: Patient Diagnosis and Class 2: Focus of Patient Evaluation and Management. Demonstrative videos of the Part II examination and an informational video about the exam day setup are available on the ABPMR website.

PART II EXAMINATION OUTLINE

Class 1: Patient Diagnosis

A. Cerebral Vascular Disease:
   1. Embolic/Thrombotic
   2. Hemorrhagic
   3. Vascular Malformation
   4. Other

B. CNS:
   1. Brain Tumor
   2. Cerebral Palsy
   3. Hypoxic Ischemic Encephalopathy
   4. Movement Disorder and Parkinson Disease
   5. Infectious or Inflammatory Disease
   6. Multiple Sclerosis
   7. Other

C. Medical Conditions Resulting in Impairment or Disability:
   1. Cancer
   2. Cardiac Rehabilitation
   3. COPD

4. Other Pulmonary Problems
5. Deconditioning
6. Immunosuppressive (HIV)
7. Organ Transplantation
8. Peripheral Vascular Disease
9. Other

D. Musculoskeletal—Occupational and Sports Injuries:
1. Acute Trauma
2. Fractures
3. Overuse Syndromes/Tendinitis
4. Strains/Sprains
5. Other

E. Musculoskeletal Disorders:
1. Amputation and Limb Deficiencies
2. Burns
3. Complex Regional Pain Syndrome
4. Fibromyalgia
5. Inflammatory Arthritis
6. Joint Replacement/Arthroplasty
7. Osteoarthritis
8. Osteoporosis
9. Other

F. Neuromuscular Disorders:
1. Hereditary Myopathies and Dystrophies
2. Inflammatory Myopathies
3. Focal and Entrapment Neuropathies
4. Hereditary Neuropathy
5. Infectious or Inflammatory Neuropathy
6. Metabolic Neuropathy
7. Plexus Lesions
8. Polyneuropathies
9. Motor Neuron Disorders
10. Neuromuscular Transmission Disorders
11. Other

G. Spinal Cord Injury:
1. Infectious and Inflammatory Disease
2. Meningomyelocele and Neural Tube Defects
3. Spondylotic Myelopathy
4. Toxic/Metabolic Conditions
5. Traumatic
6. Vascular Disorders
7. Other

H. Spine Disorders and Radiculopathy:
1. Cervical Radiculopathy
2. Thoracic Radiculopathy
3. Lumbosacral Radiculopathy
4. Degenerative Disk Disease
5. Low Back Pain
6. Spondylosis and Spondylolisthesis
7. Other

I. Traumatic Brain Injury:
1. Mild
2. Moderate/Severe
3. Other

Class 2: Focus of Patient Evaluation and Management

A. Acute Pain Management
B. Chronic Pain Management
C. Cardiovascular Impairments
D. Cognitive and Language Impairments
E. Complications of Primary Diagnosis
F. Electrodiagnostic Evaluation
G. Gastrointestinal Impairments
H. Genitourinary Impairments
I. Geriatric Rehabilitation
J. Metabolic and Nutrition Conditions
K. Musculoskeletal Impairments
L. Neurological Impairments
M. Pediatric Rehabilitation
N. Pressure Ulcers and Other Skin Conditions
O. Prevention of Impairments and Disabilities
P. Psychological and Neurobehavioral Impairments
Q. Pulmonary Impairments
R. Rehabilitative Management
    1. Vocational Rehabilitation (Return to Work, etc.)
    2. Prosthetics/Orthotics (Prescription, etc.)
    3. Durable Medical Equipment
    4. Treatment Planning (Physical Therapy, Occupational Therapy, Modalities, Activities of Daily Living [ADLs], etc.)
S. Sexual Dysfunction
T. Soft Tissue Conditions and Lymphedema
U. Other or Multiple Complications

EXAMINATION RESULTS

Official notification of the availability of examination results is sent by email 6 to 8 weeks after an examination is administered. Results and score reports are viewable by signing in to the individual candidate's account on the ABPMR website. In the interest of maintaining confidentiality of candidate information, examination results are not given over the telephone, via fax, or email.

THE CERTIFICATE

Upon approval of the application and the candidate's successful completion of the examinations, the ABPMR will grant a time-limited certificate to the effect that the candidate has met the requirements of the ABPMR. The recipient of a certificate will be known as a diplomate, or a certificant, of the ABPMR. The candidate will also be enrolled in the Continuing Certification (CC) program. The CC requirements vary, depending on the year of certification.

If the Board Certification was issued in 2020 or later, the diplomate will be on a 5-year continuing certification cycle. For those diplomates with certificates issued between 2012 through 2019, they are on a 10-year continuing certification cycle. Those diplomates with certificates prior to 2012 are also on a 10-year continuing certification cycle, however with some differences in requirements. CC procedures and requirements are described briefly in the following section and in-depth on the ABPMR website. Certificates issued prior to 1993 have no time-limited stipulations. However, holders of these pre-1993 certificates may voluntarily participate in the CC program.

Residents entering a training program must be aware that time-limited certification for PM&R began in 1993 for all diplomates certified thereafter.

PREPARATION FOR THE TEST

The ABPMR website describes the computer testing process and provides further information regarding how to prepare for the test. All candidates should read and understand the testing process, including ABPMR policies, as well as testing policies of the computer-based testing center.

Training during medical school forms the foundation on which advanced clinical knowledge is accumulated during residency training. However, the serious preparation for the examination actually starts at the beginning of the residency training in PM&R. Most candidates will require a minimum of 6 to 8 months of intense preparation for the examination. "Cramming" just before the examination is counterproductive and not recommended. Some of the methods for preparation for the Board examination are described later. Additionally, each candidate may develop their own system.

It is essential that each candidate study a standard textbook of PM&R from beginning to end. Any of the standard textbooks of PM&R should provide a good basic knowledge base in all areas of PM&R. Ideally, the candidate should read one good textbook and not jump from one to another, except for reading certain chapters that are outstanding in a particular textbook. This book and similar board review syllabi are excellent tools for brushing up on important Board-relevant information several weeks to months before the examination. They, however, cannot take the place of comprehensive textbooks of PM&R. This book is designed as a study guide rather than a comprehensive textbook of PM&R. Therefore, it should not be used as the sole source of medical information for the examination.

HELPFUL RESOURCES

In June of 2015, the ABPMR released "Part I Practice Questions," which contains 100 practice questions and is available on the ABPMR website. These are extremely valuable for obtaining practice in answering multiple-choice questions. These annual exam questions are available in print format from the American Academy of Physical Medicine and Rehabilitation (AAPM&R). These questions are not used on the Board exams, but serve as a means to assess your knowledge on a range of PM&R topics. These study guides are available on the AAPM&R website at www.aapmr.org.

Formation of study groups, three to five candidates per group, permits study of different textbooks and review articles in journals. It is important that the group meet regularly, and each candidate should be assigned reading materials. Selected review papers and state-of-the-art articles on common and important topics in PM&R should be included in the study materials.

Indiscriminate reading of articles from many journals should be avoided. In any case, most candidates who begin preparation 6 to 8 months before the examination will not find time for extensive study of journal materials. Notes and other materials the candidates have gathered during their residency training are also good sources of information. These clinical "pearls" gathered from mentors will be of help in remembering certain important points.

Certain diseases, many peculiar and uncommon, are eminently "Board-eligible," meaning that they may appear in the Board examinations more frequently than in clinical practice. Most of these are covered in this book. Several formulas and points should be memorized (such as Target Heart Rate). Most significantly, the clinical training obtained and the regular study habits formed during residency training are the most important aspects of preparation for the examination. Review courses are also available if desired.

DAY OF THE EXAMINATION

Plan to arrive at the testing center at least 30 minutes prior to your exam start time. Prior to starting the exam, you will be asked to present two forms of identification, sign the ABPMR rules, and complete both the palm vein scan and the pocket check. You will be provided a locker in which you may store your personal items. An erasable note board and marker will be provided for use during the exam.

During the exam, adequate time is allowed to read and answer all the questions. Therefore, there is no need to rush or become anxious. You should watch the time to ensure that you are at least halfway through the examination when half of the time has elapsed. Start by answering the first question, and continue sequentially (do not skip too many). Do not be alarmed by lengthy questions; look for the question's salient points. When faced with a confusing question, do not become distracted by that question. Mark it so you can find it later, go to the next question, and then come back to the unanswered ones at the end. Extremely lengthy stem statements or case presentations are apparently intended to test the candidate's ability to separate the essential from the unnecessary or unimportant information.

Some candidates may fail the examination despite the possession of an immense amount of knowledge and the clinical competence necessary to pass the examination. Their failure to pass the examination may be caused by the lack of ability to understand or interpret the questions properly. The ability to understand the nuances of the question format is sometimes referred to as "boardsmanship." Intelligent interpretation of the questions is very important for candidates who are not well versed in the format of multiple-choice questions. It is very important to read the final sentence (that appears just before the multiple answers) several times to understand how an answer should be selected. For example, the question may ask you to select the *correct* or *incorrect* answer. Nevertheless, it is advisable to recheck the question format before selecting the correct answer. It is important to read each answer option thoroughly through to the end. Occasionally, a response may be only partially correct. Watch for qualifiers such as "next," "immediately," or "initially." Another hint for selecting the correct answer is to avoid answers that contain absolute or very restrictive words such as "always," "never," or "must." Another means to ensure that you know the correct answer is to cover the answers before tackling the question. Read each question and then try to think of the answer before looking at the list of potential answers. Assume you have been given all the necessary information to answer the question. If the answer you had formulated is not among the list of answers provided, you may have interpreted the question incorrectly. When a patient's case is presented, write down the diagnosis before looking at the list of answers. It will be reassuring to realize (particularly if your diagnosis is supported by the answers) that you are on the "right track." If you do not know the answer to the question, very often you are able to rule out one or several answer options and improve your odds at guessing.

Candidates are well advised to use the basic fund of knowledge accumulated from clinical experience and reading to solve the questions. Approaching the questions as "real-life" encounters with patients is far better than trying to second-guess the examiners or trying to analyze whether the question is tricky. There is no reason for the ABPMR to trick the candidates into choosing the wrong answers.

It is better not to discuss the questions or answers (after the examination) with other candidates. Such discussions usually cause more consternation, although some candidates may derive a false sense of having performed well in the examination. In any case, candidates are bound by their oath to the ABPMR not to discuss or disseminate the questions.

CONTINUING CERTIFICATION

It is the applicant's responsibility to seek information concerning the current requirements of recertification in PM&R. The most current requirements supersede any prior requirements and are applicable to each candidate for recertification.

Prior to 2020, the ABPMR issued time-limited certificates that are valid for 10 years. However, beginning in 2020, the ABPMR began issuing time-limited certificates that are valid for 5 years. Importantly, the ABPMR is honoring all 10-year certificates. To maintain certification beyond the time-limited period, diplomates certified in 1993 and thereafter, as well as those holding a subspecialty certificate, must participate in the CC program. The intent of the initial certification and subsequent CC process is to provide assurance to the public that a certified medical specialist has successfully completed an approved educational program and an evaluation, including an examination process, designed to assess the knowledge, experience, and skills requisite to the provision of high-quality patient care in that specialty.

CC COMPONENTS

The CC program is based on documentation of individual participation in four areas: (a) professionalism, (b) lifelong learning, (c) assessment, and (d) improving health and health care (quality Improvement). Through these components, the CC addresses five core competencies—patient care, lifelong learning, maintenance of knowledge and skills, certification of the specialty, and continuous improvement.

CC REQUIREMENTS

COMPONENT I: PROFESSIONALISM

In order to maintain ABPMR certification, diplomates must adhere to the ABPMR definition of professionalism and must hold a current, valid, and unrestricted license to practice medicine. Failure to retain a valid, unrestricted license will result in the loss of ABPMR certification. The full ABPMR Definition of Professionalism is available on the ABPMR website. In the event that a diplomate's license to practice medicine is revoked, suspended, or surrendered, ABPMR certification will be simultaneously revoked. Further details regarding professional standing are outlined in the ABPMR Policy on Licensure, which is also available on the ABPMR website.

COMPONENT II: LIFELONG LEARNING

Continuing Medical Education (CME) Requirement

Starting in 2020, the ABPMR began including Self-Assessment CME credits as part of the total CME credit requirement. A list of approved ABPMR self-assessment options can be found on the ABPMR website. Diplomates are encouraged to complete and report Category 1 CME credits and Self-Assessment CME credits annually. Diplomates with time-limited certificates issued between 2012 and 2019 must complete and report 150 Category 1 CME credits, of which 40 must be Self-Assessment (SA) credits, in years 1 to 5 and again in years 6 to 10 of their CC cycle, for a total of 300 Category 1 CME credits, of which at least 80 credits must be SA-CME. Diplomates with time-limited certificates issued in 2020 and beyond must complete and report 150 Category 1 CME credits, of which 50 credits must be SA-CME, in their 5-year CC cycle.

Certificates for Category 1 CME activities should be retained by the diplomate in the event that the Board requests verification. A minimum of 50% of the 300 total CME credits must be specifically related to the specialty of PM&R and/or its subspecialties.

Category 1 credit involves activities designated by an accredited provider. A minimum of 300 credits must be met by the following types of CME experiences:

- CME programs of universities, hospitals, organizations, and institutions accredited by the Accreditation Council for Continuing Medical Education (ACCME).
- CME activities offered by other accrediting organizations such as the American Medical Association (AMA), the AAPM&R, the Association of Academic Physiatrists (AAP), or the American Association of Neuromuscular and Electrodiagnostic Medicine (AANEM).
- Category 1A and 2A credits from the American Osteopathic Association (AOA).

Category 2 credits may be reported for tracking purposes only and do not count toward the 300-credit minimum.

Self-Assessment Requirement

Diplomates with time-limited certificates issued before 2012 are required to complete four ABPMR-approved self-assessment activities during the 10-year MOC cycle. Diplomates with time-limited certificates issued in 2012 and beyond must complete Self-Assessment CME as described in the prior section. A list of approved ABPMR self-assessment options can be found on the ABPMR website.

COMPONENT III: ASSESSMENT

Longitudinal Assessment for PM&R (LA-PM&R)

Beginning in 2020, ABPMR diplomates began longitudinal assessment using the LA-PM&R, which replaced the MOC® Examination. The LA-PM&R is administered through the LA-PM&R Platform on the ABPMR website. Completion of the LA-PM&R is required on a quarterly basis throughout years 1 to 4 of the diplomate's 5-year certification period, for a total of 16 separate assessments, with a summative assessment in the fifth year. Participation in the LA-PM&R during year 5 is optional but allows for continued learning and additional SA-CME.

LA-PM&R Format

The LA-PM&R employs three different types of questions, which appear in different quantities depending on the quarter in which the LA-PM&R is taken: multiple-choice, reattempted multiple-choice, and article-based. In the first quarter, the LA-PM&R contains 20 multiple-choice questions. In the second quarter, it contains 15 new multiple-choice questions plus any questions answered incorrectly from the first quarter, called reattempt questions. In a similar fashion, the third quarter LA-PM&R contains 15 new questions plus any reattempt questions from the second quarter. In the fourth quarter, the LA-PM&R contains 10 article-based questions relating to five PM&R relevant articles (two questions per article) plus any reattempt questions from the third quarter.

Three minutes are allotted per multiple-choice and reattempt question, while 15 minutes are allotted per article-based question. It is important to note that correctly answered reattempt questions are given full credit: no penalty is given to questions previously answered incorrectly and subsequently answered correctly on reattempt. Further, if an article-based question is answered incorrectly, a rationale relating to the article is given and the question may be immediately reattempted for full credit. Diplomates who answer all questions each quarter may claim three SA-CME credits per quarter, for a total of 12 SA-CME per year. Each quarterly LA may be completed at any time before the respective deadlines:

Q1: March 31
Q2: June 30
Q3: September 31
Q4: December 31

LA-PM&R Content Customization

Unlike the previous MOC® exam or the Part I and II exams, LA-PM&R does not have exact target content weights. Instead, diplomates may customize the relative number of questions from the five core content categories based on their learning goals or practice area. Diplomates may design their LA-PM&R to draw up to 50% of its questions from a single core category; however, the LA must draw a minimum of 10% of its questions from each core category. A key exception is the Pediatric Rehabilitation Medicine category, which may be excluded from the LA-PM&R entirely should the diplomate wish to do so. Diplomates choose the relative target weight of each core category at the beginning of the calendar year. Once chosen, the relative target weights of each core category will be represented on each of the respective calendar year's quarterly

assessments and may not be changed until the following calendar year. The article-based questions offer an additional level of assessment customization: 10 articles are provided at the beginning of Q4 on the LA-PM&R platform, of which the diplomate must choose five on which to be assessed. These articles are available for reading both prior to and while attempting the article-based questions. A video tutorial of LA-PM&R content customization is available on the ABPMR website. The five core categories and their relative weight ranges are listed below with each major subcategory.

- Central Nervous System Rehabilitation (10%–50%)
  - Brain Injury
  - Spinal Cord Injury
  - Stroke
- Musculoskeletal and Pain Medicine (10%–50%)
  - Arthritis
  - Acute/Chronic Trauma
  - Fractures
  - Spinal Disorders
  - Sports Medicine
- Medical Rehabilitation (10%–50%)
  - Cardiopulmonary
  - Amputation/Wounds
  - Cancer
  - Polytrauma
  - Geriatric Disorders
  - Transplant
- Neuromuscular Medicine and Electrodiagnosis (10%–50%)
  - Neuropathies
  - Myopathies
  - Motor Neuron Disease
  - Electrodiagnostic Studies
- Pediatric Rehabilitation Medicine (0%–50%)

LA-PM&R Scoring

The scores from the previous 16 quarters of LA-PM&R testing will be examined, with the lowest four quarters removed. Diplomates who have elected to not participate in up to four quarters of LA-PM&R are thus not penalized. In the fifth year of the diplomate's CC cycle, a summative assessment is made of whether the diplomate has met the performance standard of the ABPMR. As of the time of writing, the ABPMR is continuing to accumulate LA-PM&R data and, as a result, has not yet determined the final performance standard for the LA-PM&R. When determined, diplomates will be able to compare their performance on each quarterly LA-PM&R to the performance standard.

Re-Entry Exam

For individuals who do not meet the LA-PM&R performance standard, or for those whose primary certification has expired, certification must be attained by passing the PM&R Re-Entry examination. It should be noted that for diplomates who did not meet LA-PM&R standards, all other requirements for CC must still be met. The Re-Entry examination is offered once per year in the late summer/early fall and comprises 250 multiple-choice questions divided between two 3.5-hour sections. A 60-minute break is provided between the two sections. The Re-Entry exam is modeled on the difficulty and general outline of the Part I examination for initial certification. The major content domains are shared with the Part I examination but have slightly different target weights. Individuals should review the ABPMR website for a breakdown of the content domains for the Re-Entry exam.

COMPONENT IV: IMPROVING HEALTH AND HEALTH CARE (QUALITY IMPROVEMENT)

The fourth component contains various assessments designed to address improving health and health care quality improvement (IHHC QI) in practice.

Diplomates with time-limited certificates issued between 2012 and 2019 beyond must complete two ABPMR-approved quality improvement projects (one in years 1–5 and the second in years 6–10) during the 10-year CC cycle. Diplomates with time-limited certificates issued in 2020 and beyond must complete one of several IHHC QI options during their 5-year certification cycle. A list of ABPMR-approved IHHC QI project options can be found on the ABPMR website along with further details about how to submit the IHHC QI project.

CC REQUIREMENTS SUMMARY

CERTIFICATE ISSUE DATE	ACTIVITIES REQUIRED TO RECERTIFY
2012–2019	• Maintain licensure. • Pay annual program fee. • Adhere to ABPMR Principles of Professionalism. • Complete and report a minimum of 150 Category 1 CME credits (40 of which must be SA-CME) in years 1–5 and 150 Category 1 CME credits (including 40 SA-CME) in years 6–10, for a total of 300 Category 1 CME credits (including 80 SA-CME credits). • Complete two ABPMR-approved quality improvement projects (one in years 1–5 and one in years 6–10). • Participate in LA-PM&R Assessment.
2020 and beyond	• Maintain licensure. • Pay annual program fee. • Adhere to ABPMR Principles of Professionalism. • Complete and report a minimum of 150 Category 1 CME credits in years 1–5, including 50 SA-CME credits. • Participate in LA-PM&R Assessment. • Complete one ABPMR-approved IHHC QI project.

ABPMR, American Board of Physical Medicine and Rehabilitation; CME, Continuing Medical Education; IHHC QI, improving health and healthcare quality improvement; LA-PM&R, Longitudinal Assessment for PM&R; SA-CME, Self-Assessment Continuing Medical Education.

CERTIFICATE ISSUANCE

The Board will issue a 5-year time-limited certificate to each diplomate who successfully completes the CC process. Prior to receiving a certificate, diplomates must complete all CC components and pay all annual fees that are due.

Diplomates who have not completed all CC program requirements prior to the expiration date of their certificate may reinstate their diplomate status by taking the PM&R Re-Entry exam, pursuant to the ABPMR CC Reinstatement Policy.

EXAMINATION STATISTICS

TOTAL PM&R DIPLOMATES CERTIFIED AS OF 2024: 15,536

PART I: COMPUTER-BASED EXAMINATION	AUGUST 2024
Total taking exam	552
Total taking exam for the first time	498
Total first-time (pass)	429/86.1%
TOTAL FIRST-TIME (FAIL)	69/13.9%

PART II: ORAL EXAMINATION	MAY 2024
Total taking exam	517
Total taking exam for the first time	444
Total first-time (pass)	389/87.6%
TOTAL FIRST-TIME (FAIL)	55/12.4%

PM&R RE-ENTRY EXAMINATION	JANUARY 2025
Total taking exam	47
Total taking exam for the first time	41
Total first-time (pass)	40/97.6%
Total first-time (fail)	1/2.4%

Further details and current information for the certification and recertification programs can be obtained by contacting the ABPMR.

The American Board of Physical Medicine and Rehabilitation
3015 Allegro Park Lane SW
Rochester MN 55902-4139
Phone: 507-282-1776Website: www.abpmr.org
Email: office@abpmr.org

STROKE

1

Richard D. Zorowitz, MD • Edgardo Baerga, MD • Sara J. Cuccurullo, MD • Talya Fleming, MD • Stacey Jou, MD

INTRODUCTION

DEFINITION OF STROKE

- An episode of neurologic dysfunction persisting ≥24 hours or until death; causes include focal cerebral, spinal, or retinal infarction, as well as focal bleed in the brain parenchyma or ventricular system not caused by trauma, and bleed into the subarachnoid space not caused by trauma (Sacco et al., 2013).
- Transient ischemic attack (TIA) = "a transient episode of neurological dysfunction caused by focal brain, spinal cord, or retinal ischemia without acute infarction" (Easton et al., 2009).

EPIDEMIOLOGY

- Stroke is the fifth leading cause of death in the United States, with the top 6 in the following order: heart disease, cancer, COVID-19, accidents, stroke, and chronic lower respiratory diseases (Xu et al., 2022).
- The American Heart Association (AHA) estimates 795,000 strokes annually: 610,000 new cases and 185,000 recurrent cases (Benjamin et al., 2018; Tsao et al., 2023).
- There were nearly 6.8 million (about 3.0 million male and 3.8 million female) stroke survivors in the United States in 2010.
- Females (one in five) have a higher lifetime risk of stroke than males (approximately one in six; Tsao et al., 2023).
- Racial and ethnic disparities in stroke risk may persist or even increase in older adult females from underrepresented races and ethnicities (Tsao et al., 2023).
- Mortality rates were higher after an incident hemorrhagic stroke (67.9%) than after ischemic stroke (57.4%; Tsao et al., 2023).
- Age-adjusted mortality after an incident stroke decreased over time (absolute decrease of 8.1 deaths per 100 strokes after 10 years), which was attributed mainly to the decrease in mortality among those ≤65 years of age (absolute decrease of 14.2 deaths per 100 strokes after 10 years; Tsao et al., 2023).
- A 46% decline in cerebral infarcts and hemorrhages was observed from the 1950 to 1954 period to the 1975 to 1979 period (Broderick, 1993).
  - Decline was attributed to better management of hypertension (HTN) and heart disease, and a decrease in cigarette smoking.
- Incidence increased 17% from the 1975 to 1979 period to the 1980 to 1984 period (attributed to increased use of CT scan).

- From 1990 to 2019, the absolute number of incident strokes increased globally by 70.0% but the age-standardized incidence rate for total stroke decreased by 17%.
- Incidence of reported ruptured aneurysm is around 10 in every 100,000 persons per year (about 30,000 individuals per year in the United States).
- Overall, mortality from strokes has declined since the 1950s.
  - A sharp decline was noted in the 1970s, possibly related to improved diagnosis and treatment of HTN.
  - From 1999 to 2009, the annual stroke death rate decreased 36.9%, and the actual number of stroke deaths declined 22.9%.
  - From 2010 to 2020, in the United States, the actual number of stroke deaths increased 23.8% (from 129,476 to 160,264), whereas the age-adjusted stroke death rate decreased 0.8% (Tsao et al., 2023).
  - Approximately 62% of stroke deaths in 2015 occurred outside the hospital.
  - Data from the Atherosclerosis Risk in Communities (ARIC) study (1987–2011 in four U.S. cities) showed that the cumulative all-cause mortality rate after a stroke was 10.5% at 30 days, 21.2% at 1 year, 39.8% at 5 years, and 58.4% at the end of 24 years of follow-up (Benjamin et al., 2018; Tsao et al 2023).
  - In the first 30 days after stroke, 90% of deaths were due to direct effects of the brain lesion or complications of immobility resulting from the stroke (Dennis et al., 1993).
  - Between 30 days and 6 months after stroke, 44% of deaths were secondary to complications or direct effects of the first stroke (Dennis et al., 1993).
  - After 6 months, the most likely cause of death is cardiovascular (nonstroke) or sudden death (Dennis et al., 1993).
  - The age-adjusted death rate for stroke in 2020 in the United States for non-Hispanic Black females and males was higher than for Hispanic females and males, and non-Hispanic White, Asian, American Indian, or Alaska Native females and males (Tsao et al., 2023).
- Risk of stroke following TIA:
  - Approximately 12% of all strokes are heralded by a TIA. The prevalence of TIA in men 65 to 69 years is 2.7%, and 3.6% in men 75 to 79 years (Kleindorfer et al., 2005; Lisabeth et al., 2004).
  - The short-term risk of stroke after TIA is 1.2% at 2 days and 7.4% at 90 days (Tsao et al., 2023).
  - Individuals who have a TIA and survive the initial high-risk period have a 10-year stroke risk of roughly 19% and a combined 10-year stroke, myocardial infarction (MI), or vascular death risk of 43% (4% per year; Benjamin et al., 2018).

🕮 RISK FACTORS (Stewart, 1999)

Nonmodifiable Risk Factors

- Age—the single most important risk factor for stroke worldwide. After age 55, incidence increases for both males and females.
  - Risk more than doubles each decade after age 55.
- Sex (male > female)
- Race (African Americans 2× > Caucasians > Asians)
- Family history of stroke

Modifiable (Treatable) Risk Factors

- 87% of stroke risk can be attributed to modifiable risk factors.
- **HTN**: Most important modifiable risk factor for both ischemic and hemorrhagic stroke (sevenfold increased risk). Lower rates of recurrent stroke with lower blood pressures (BPs). Most recently, the BP-reduction component of the Secondary Prevention of Small Subcortical Strokes (SPS3) trial showed that targeting a systolic BP (SBP) <130 mmHg (compared with 130–149 mmHg) was likely to reduce stroke by 20% and intracerebral hemorrhage (ICH) by two-thirds (Tsao et al., 2023).

- **History of TIA/prior stroke**: Approximately 5% of patients with TIA will develop a stroke within 1 month and approximately 14% within 1 year if untreated. After a TIA, the 90-day risk of stroke is 3% to 17.3% and is highest within the first 30 days (Coull et al., 2004; Johnston et al., 2003).
- **Heart disease**:
  - Congestive heart failure (CHF) and coronary artery disease (CAD) increase risk by twofold.
  - Valvular heart disease and arrhythmias increase risk of embolic stroke.
- 🕮 **Atrial fibrillation (AFib)**: Dominant risk factor for stroke, independently increasing risk fivefold throughout all ages (Benjamin et al., 2018)
  - The percentage of strokes attributable to AFib increases from 1.5% at 50 to 59 years of age to 23.5% at 80 to 89 years of age (Benjamin et al., 2018).
  - AFib increased with age in the stroke population (2% in age <50 years old, 15% in the 70s, 28% in the 80s, and 40% in >90 years old; Jørgensen et al., 1996).
  - AFib was associated with increased age, CAD, previous stroke, and increased SBP but not with gender, diabetes mellitus, or previous TIA (Jørgensen et al., 1996).
- **Diabetes**: Twofold increase in risk. Duration of diabetes of > 3 years was associated with an increased rate of ischemic stroke compared with a duration of <3 years (Benjamin et al., 2018).
- **Cigarette smoking**: Cigarette smoking is a risk factor for ischemic stroke and subarachnoid hemorrhage (SAH). Current smokers have a two to four times increased risk of stroke compared with nonsmokers or those who have quit for >10 years (Benjamin et al., 2018).
- **Carotid stenosis (and carotid bruit)**: Carotid endarterectomy (CEA) is of some benefit to prevent stroke in patients with 50% to 69% symptomatic stenosis (absolute risk reduction 4.6%) and highly beneficial in patients with 70% to 99% stenosis (absolute risk reduction 16.0%).
- **Ethyl alcohol (ETOH) abuse/cocaine use**: <2 drinks per day relative risk 0.51; >7 drinks per day relative risk 2.96 (Sacco et al., 1999).
- **High-dose estrogens (birth control pills)**: Considerable increased risk when linked with cigarette smoking.
- **Systemic diseases associated with hypercoagulable states**:
  - Elevated red blood cell (RBC) count, hematocrit, fibrinogen
  - Protein S and C deficiencies
  - Sickle cell anemia
  - Cancer
- **Hyperlipidemia**: Several clinical trials have shown a reduction in stroke with use of cholesterol-reducing agents (approximately 30% reduction risk of stroke with use of 3-hydroxy-3-methylglutaryl-coenzyme A [HMG-CoA] reductase inhibitors).
  - Low-density lipoprotein-cholesterol (LDL-C) was associated with an increased risk of stroke, and LDL-C may have a stronger association for the large artery atherosclerotic subtype (Benjamin et al., 2018).
- **Migraine headaches (HAs)**
- **Sleep apnea**
- **Patent foramen ovale (PFO)**
- **Physical activity**: Multiple studies have shown a significant association between sedentary time (i.e., seated rest) duration and risk of cardiovascular disease and stroke, independent of physical activity levels (Benjamin et al., 2018).
- **Nutrition**: Compliance with a Mediterranean-style diet that was higher in nuts and olive oil was associated with a reduced incidence of stroke (Benjamin et al., 2018).

Other Risk Factors (Benjamin et al., 2018)

- **Geographic location:** Higher risk of stroke is found in the southeastern United States than in other areas—the so-called "stroke belt" states (North Carolina, South Carolina, Georgia, Tennessee, Mississippi, Alabama, Louisiana, and Arkansas).
- **Socioeconomic factors:** There is some evidence that strokes are more common in people with low-income status than among more affluent people.

Select Stroke Risk Factors

STROKE RISK FACTOR	INCREASE IN STROKE RISK
Age over 55 years old	Doubles risk every 10 years
Hypertension	7×
Congestive heart failure	2×
Atrial fibrillation	5×
Coronary artery disease	2×
Diabetes	2×
Smoking	2–4×

BASIC NEUROANATOMIC REVIEW OF THE MAJOR VESSELS INVOLVED IN STROKE (FIGURES 1–1 TO 1–4)

1. The **middle cerebral artery (MCA)** supplies most of the lateral aspect of the hemisphere.
2. The **anterior cerebral artery (ACA)** supplies the medial aspect of the hemisphere from the lamina terminalis to the cuneus.
3. The **posterior cerebral artery (PCA)** supplies the posterior inferior surface of the temporal lobe and the visual cortex.

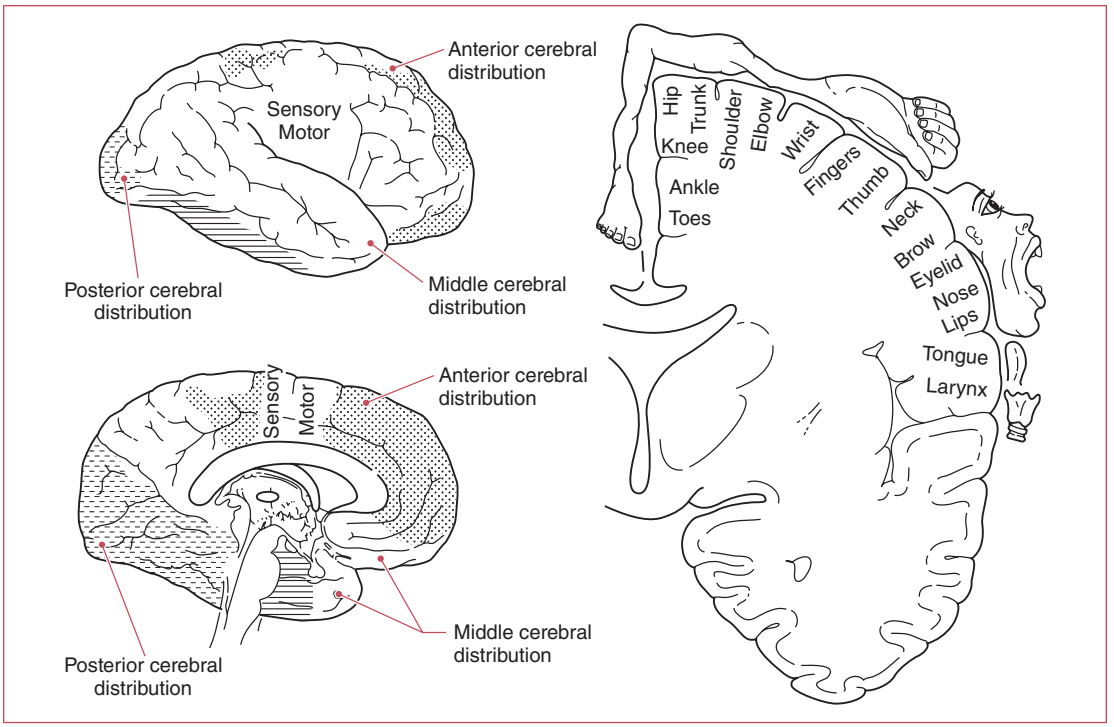


FIGURE 1–1 Major vascular supply to the brain and functional diagram of the motor strip. It is evident that the lower limb extremity motor strip is in ACA distribution, while the upper extremity motor strip is supplied by the MCA. ACA, anterior cerebral artery; MCA, middle cerebral artery.
Source: From Rosen P. *Emergency Medicine—Stroke*. 3rd ed. Mosby; 1992, with permission.

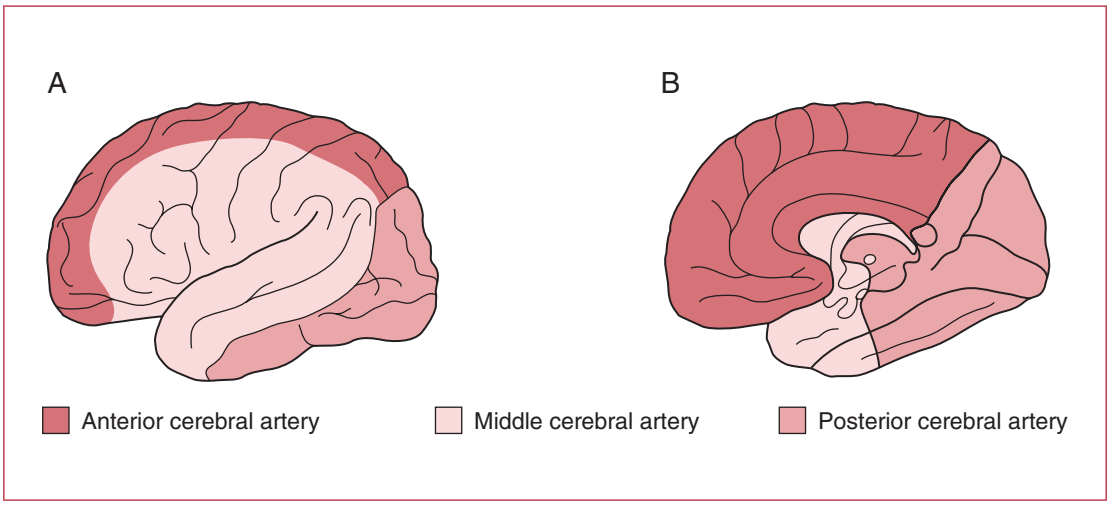


FIGURE 1–2 The three cerebral arteries' cortical territories. (A) Lateral aspect of the hemisphere. (B) Medial and inferior aspects of the hemisphere.

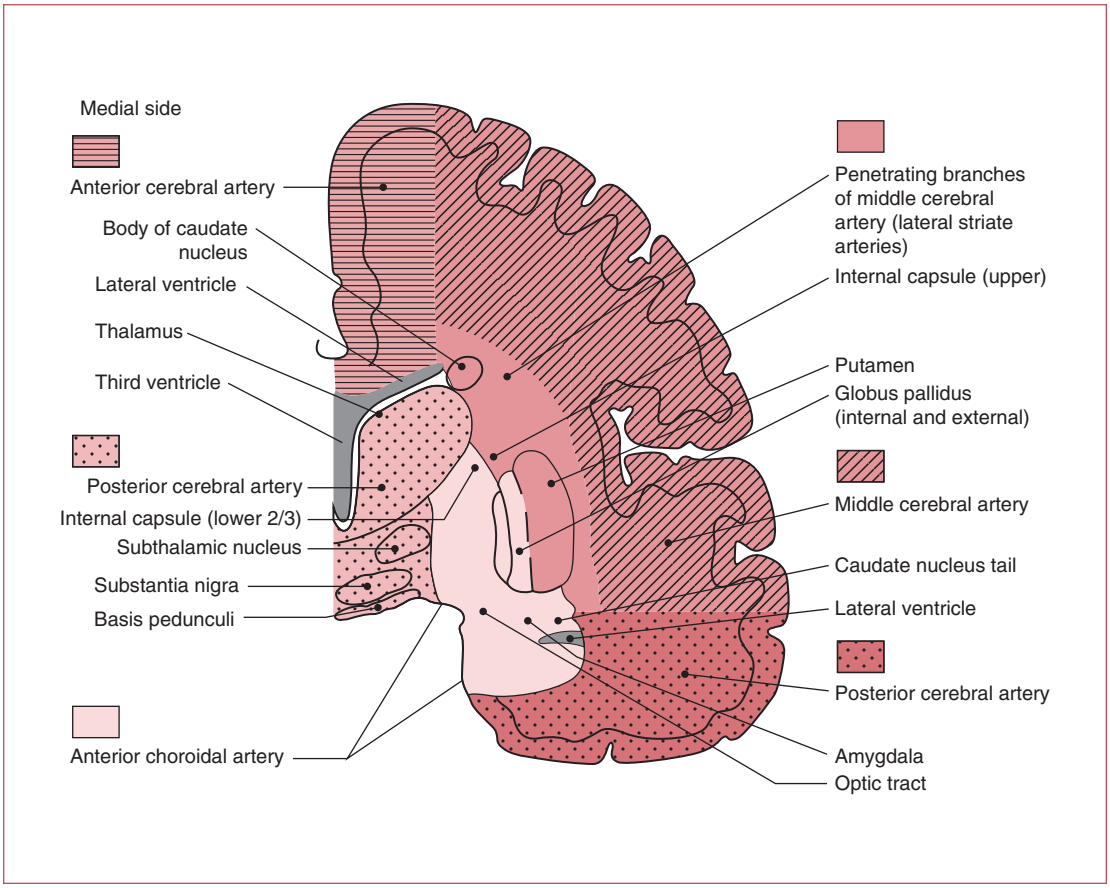


FIGURE 1–3 Major vascular territories are shown in this schematic drawing of a coronal section through the cerebral hemisphere at the level of the thalamus and the internal capsule.

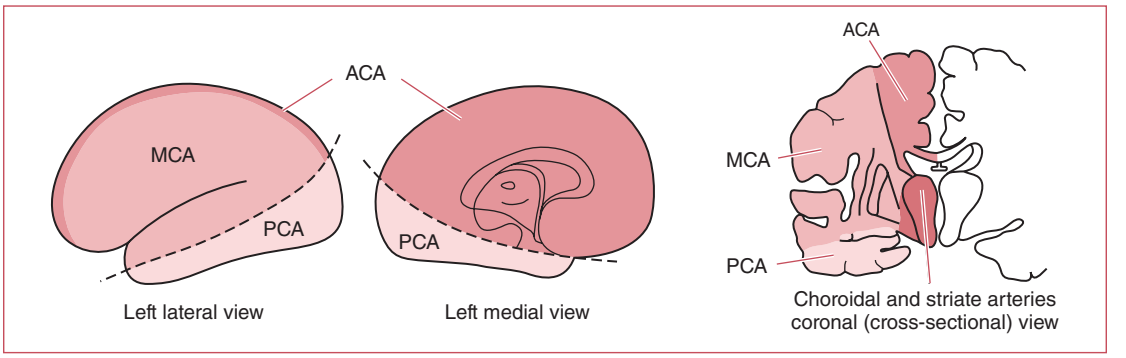


FIGURE 1–4 The cerebral blood circulation.
ACA, anterior cerebral artery; MCA, middle cerebral artery; PCA, posterior cerebral artery.

CEREBROSPINAL FLUID CIRCULATION (FIGURE 1–5)

1. 🕮 Cerebrospinal fluid (CSF) is produced in the brain largely by modified ependymal cells in the choroid plexus in the lateral, third, and fourth ventricles, with the remainder formed around blood vessels and along ventricular walls.
2. 🕮 CSF circulates from the lateral ventricles to the foramina of Monro (interventricular foramina), third ventricle, aqueduct of Sylvius (cerebral aqueduct), fourth ventricle, foramen of Magendie (median aperture) and foramina of Luschka (lateral apertures), and subarachnoid space over the brain and spinal cord.
3. Hemorrhage can cause occlusion of the foramina, resulting in hydrocephalus.

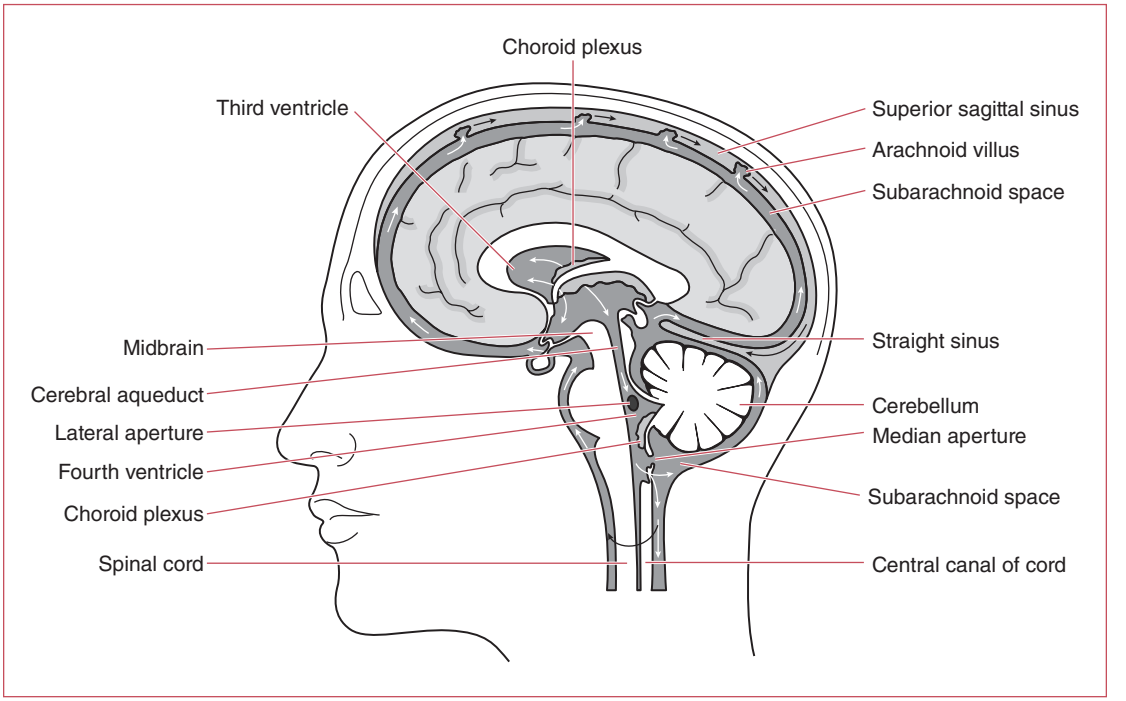


FIGURE 1–5 CSF circulation.
CSF, cerebrospinal fluid.

TYPES OF STROKE (TABLE 1–1)

TABLE 1–1 Types of Stroke

	ISCHEMIC (87%)			HEMORRHAGIC (13%)	
	Thrombotic	Embolic	Lacunar	ICH—Hypertensive	SAH—Ruptured Aneurysm
Frequency (%)	48	26	13	10	3
Factors associated with onset	Occurs during sleep	Occurs while awake		90% of cases occurs when patient is calm and unstressed; Blacks > Whites	Occurs during activity (often strenuous activity)
Major causes/etiology	Perfusion failure distal to site of severe stenosis or occlusion of major vessels	Due mainly to cardiac source	Small lesions (<15–20 mm) seen mainly in: • Putamen • Pons • Thalamus • Caudate • Internal capsule/corona radiata	Hypertension	From ruptured aneurysms and vascular malformations
Presentation	Slow, gradual, progressive onset of deficit	Sudden, immediate deficit (seizures may occur)	Abrupt or gradual onset	Gradual onset (minutes to days) or sudden onset of local neurologic deficits	Sudden onset
Association with TIA	50% with preceding TIA (50% occurring in the same vascular territory of the preceding TIA)	TIA less common than in thrombotic; 11% with preceding TIA	23% with preceding TIA	8% with preceding TIA	7% with preceding TIA

ICH, intracerebral hemorrhage; SAH, subarachnoid hemorrhage; TIA, transient ischemic attack.

ISCHEMIC STROKES

THROMBOTIC STROKES (LARGE ARTERY THROMBOSIS)

- Accounts for 48% of all strokes.
- Usually occurs during sleep (patient often awakens unaware of deficits).
- May have "stuttering," intermittent progression of neurologic deficits, or be slowly progressive (over 24–48 hours).
- Profound loss of consciousness (LOC) is rare except when area of infarction is large or when brainstem is involved.
- Neurologic deficit varies according to cerebral territory affected.

- Perfusion failure distal to site of severe stenosis or occlusion of major vessels.
- Emboli from incompletely thrombosed artery may precipitate an abrupt deficit. May have embolism from extracranial arteries affected by stenosis or ulcer.

EMBOLIC STROKES

- Accounts for 26% of all strokes.
- Immediate onset of neurologic deficits.
- Usually occurs during waking hours.
- Seizures may occur at onset of stroke.
- Most commonly due to cardiac source: Mural thrombi and platelet aggregates.
- Emboli most commonly originate from cardiac thrombus caused by AFib. Also occur in rheumatic heart disease (e.g., mitral stenosis), post-MI, and vegetations on heart valves in bacterial or marantic endocarditis or prosthetic heart valves.
  - From clots that develop after open-heart surgery or atheromas in neck arteries or in the aortic arch.
  - Emboli may dislodge spontaneously or after invasive cardiovascular procedures (e.g., cardiac catheterization).
  - 📖 75% of cardiogenic emboli go to the brain.
- Occasionally, embolus may consist of fat (from fractured long bones), air (in decompression sickness), or venous clot that passes through a PFO with shunt (paradoxical embolus).
- Rarely, a subclavian artery thrombosis may embolize to the vertebral artery or its branches.

LACUNAR STROKES (TABLE 1–4)

- Accounts for 13% of all strokes.
- Onset may be abrupt or gradual: Up to 30% develop slowly over or up to 36 hours.
- Lacunes are small infarcts (<15 mm) seen in the putamen, pons, thalamus, caudate, and internal capsule.
- They are due to occlusive arteriolar or small artery disease (occlusion of deep penetrating branches of large vessels).
- Occlusion occurs in small arteries of 50 to 200 mm in diameter.
- Strong correlation with HTN (up to 81%); also associated with microatheroma, microembolism, or rarely arteritis.
- CT shows lesion in about two-thirds of cases (MRI may be more sensitive).
- Often relatively pure syndromes (motor, sensory); discussed later.
- Absence of higher cortical function involvement (language, praxis, nondominant hemisphere syndrome, and vision).

Neuroanatomic Locations of Ischemic Stroke (Ropper & Samuels, 2009)

1. Anterior Circulation

INTERNAL CAROTID ARTERY (FIGURE 1–6)

- The most variable syndrome. Occlusion occurs most frequently in the first part of the internal carotid artery (ICA) immediately beyond the carotid bifurcation. ICA occlusions are often asymptomatic (30%–40% of cases).
- *Ocular infarction*: Embolic occlusion of either retinal branch or central retinal artery.
- *Transient monocular blindness (amaurosis fugax)*: The ICA nourishes the optic nerve and retina, as well as the brain. Transient monocular blindness occurs prior to onset of stroke in approximately 25% of cases of ICA occlusion. Central retinal artery ischemia is very rare because of collateral supply.
- *Cerebral infarction*: Variable presentation with complete ICA occlusion; from no symptoms (if good collateral circulation exists) to severe, massive infarction in ACA and MCA distributions.

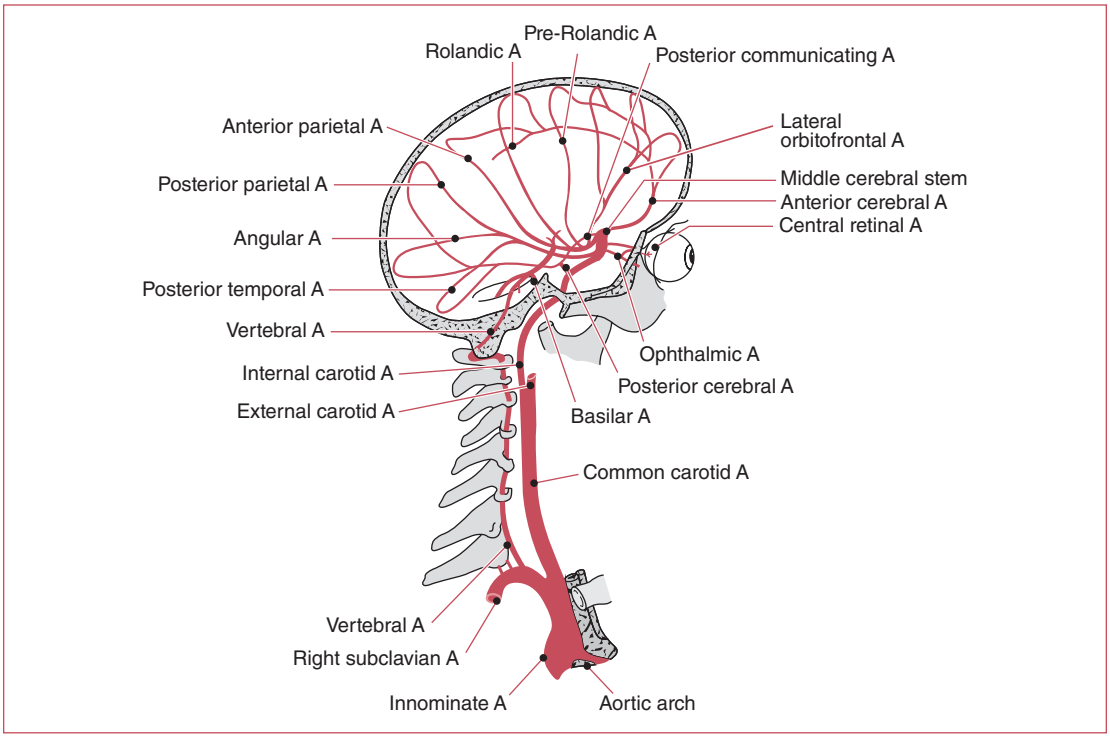


FIGURE 1–6 Arterial anatomy of major vessels on the right side carrying blood from the heart to the brain. Note location and course of the internal carotid artery.
A, artery.

- Distal ICA occlusion affects part or all of the ipsilateral MCA territory and, when the anterior communicating artery is small, the ipsilateral ACA territory. Patients will present with contralateral motor and/or sensory symptoms.

MIDDLE CEREBRAL ARTERY (FIGURES 1–7A AND 1–7B)

Occlusion occurs at the stem of the MCA or at one of the two main divisions (the *superior* or *inferior* division of the M4 segment) of the artery in the sylvian fissure.

📖 *MIDDLE CEREBRAL ARTERY SEGMENTS*

- ***Horizontal M1 Segment***
  - Branch from the ICA, runs in sphenoidal compartment of sylvian fissure, gives off lateral lenticulostriate arteries going to basal ganglia and internal capsule. May provide early cortical branch to temporal lobe (Medrano-Martorell et al., 2021; Ulm et al., 2008).
- ***Sylvian M2 Segment***
  - From genu at limen insula, runs in insular compartment of the sylvian fissure. Branches supply the inferolateral frontal lobe, temporal lobe, insular cortex, and parietal lobe (Medrano-Martorell et al., 2021; Ulm et al., 2008).
- ***Opercular M3 Segment***
  - Runs along the frontoparietal and temporal opercula to the superficial edge of sylvian fissure. Branches supply the lateral cerebral cortex (Medrano-Martorell et al., 2021; Ulm et al., 2008).
- ***Cortical M4 Segment, Superior Division of the MCA***
  - The superior division of MCA supplies the Rolandic and pre-Rolandic areas.
  - 📖 Most common cause of occlusion of the superior division of MCA is an embolus.
    - Sensory and motor deficits on contralateral face and arm > leg.
  - Head and eyes deviated toward side of infarct.
  - With left side lesion (dominant hemisphere)—global aphasia initially, then turns into Broca's aphasia (motor speech disorder).

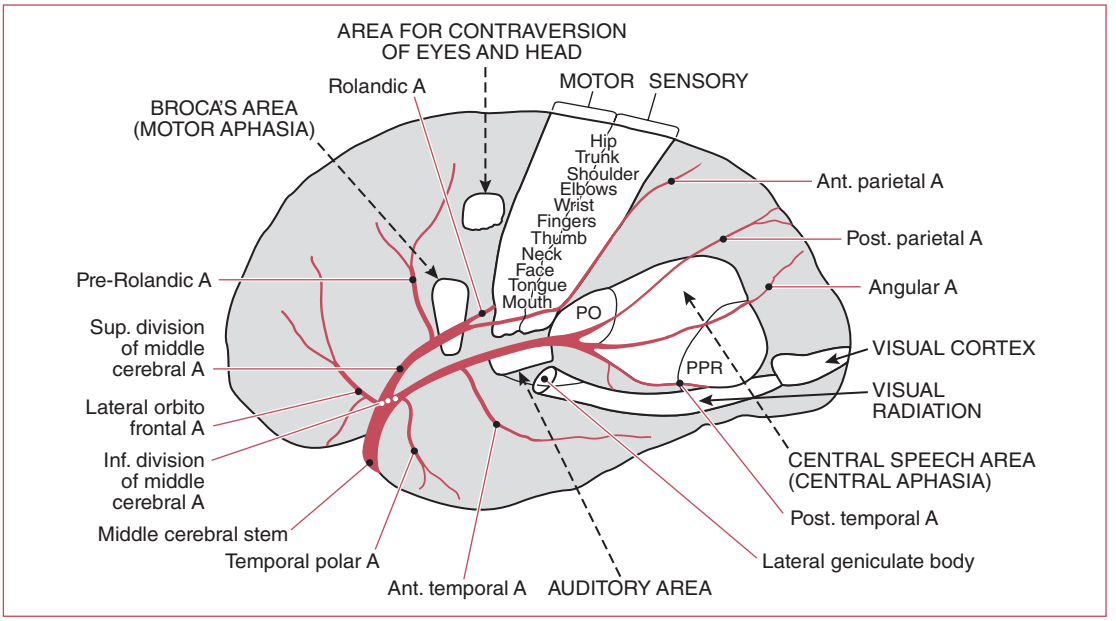


FIGURE 1–7A The distribution of the middle cerebral artery (MCA) on the lateral aspect of the cerebral hemisphere. Principal regions of cerebral localization are noted.
A, artery; Ant., anterior; Inf., inferior; PO, parietal operculum (conduction aphasia); Post., posterior; PPR, posterior parietal region (alexia with agraphia); Sup., superior.

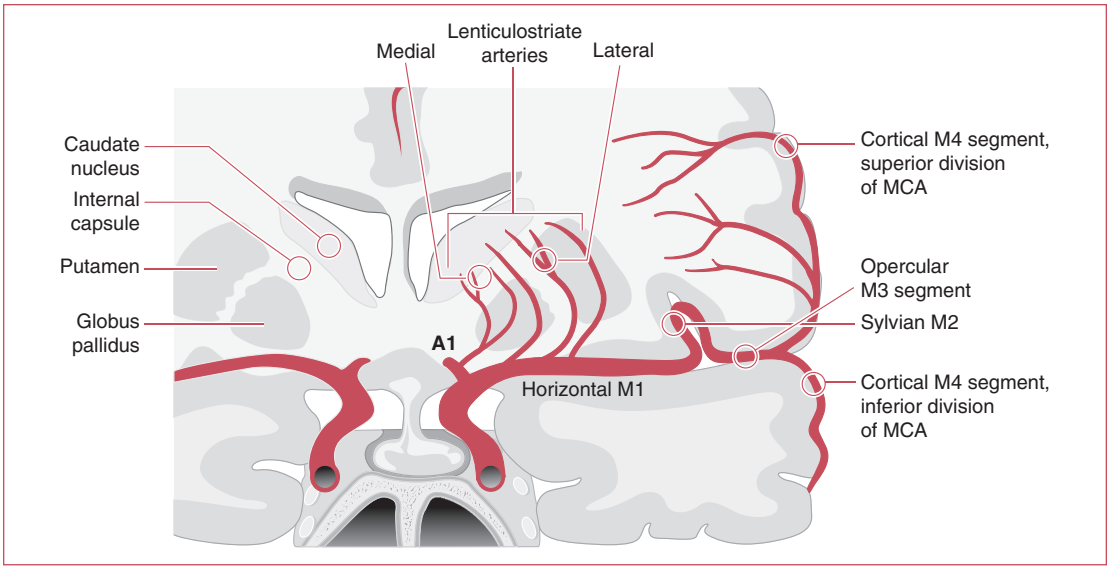


FIGURE 1–7B The horizontal M1 segment gives rise to the lateral lenticulostriate arteries, which supply the globus pallidus, putamen, and posterior limb of the internal capsule. The sylvian M2 segment supplies the temporal lobe, insular cortex, parietal lobe, and inferolateral frontal lobe. The opercular M3 segment supplies the lateral cerebral cortex. The cortical M4 segments supply the Rolandic and pre-Rolandic areas (superior division), and the lateral temporal and inferior parietal lobes (inferior division).
MCA, middle cerebral artery.

- Right side lesion (nondominant hemisphere)—deficits on spatial perception, hemineglect, constructional apraxia, and dressing apraxia.
- Muscle tone usually decreased initially and gradually increases over days or weeks to spasticity.
- Transient LOC is uncommon.

- *Cortical M4 Segment, Inferior Division of the MCA*
  - The inferior division of the MCA is the blood supply to the lateral temporal and inferior parietal lobes.
  - With lesion on either side—superior quadrantanopia or homonymous hemianopsia.
  - Left side lesion → Wernicke's aphasia.
  - Right side lesion → left visual neglect.

ANTERIOR CEREBRAL ARTERY (FIGURE 1–8)

If occlusion is at the stem of the ACA proximal to its connection with the anterior communicating artery, it is usually well-tolerated because adequate collateral circulation comes from the contralateral ACA.

- Occlusion of one ACA **distal** to an anterior communicating artery results in:
  - 🕮 Contralateral weakness and sensory loss, affecting mainly the distal contralateral leg (foot/leg more affected than thigh)
  - Mild or no involvement of upper extremity (UE)
  - Head and eyes may be deviated toward side of lesion acutely.
  - 🕮 Urinary incontinence with contralateral grasp reflex and paratonic rigidity (Gegenhalten) may be present.
  - May produce transcortical motor aphasia if left side is affected
  - 🕮 Disturbances in gait and stance = gait apraxia
- 🕮 If both ACAs arise from one stem, major disturbances occur with infarction occurring at the medial aspects of both cerebral hemispheres, resulting in aphasia, paraplegia, incontinence, and frontal lobe/personality dysfunction (e.g., emotional instability, disinhibition, and apathy).

2. Posterior Circulation: Vertebrobasilar Arteries and Posterior Cerebral Arteries

POSTERIOR CEREBRAL ARTERY (FIGURE 1–8)

- Occlusion of PCA can produce a variety of clinical effects because it supplies the upper brainstem and the inferior parts of the temporal lobe, as well as the medial parts of the occipital lobe.
- The particular area of occlusion varies for PCA because the anatomy varies.

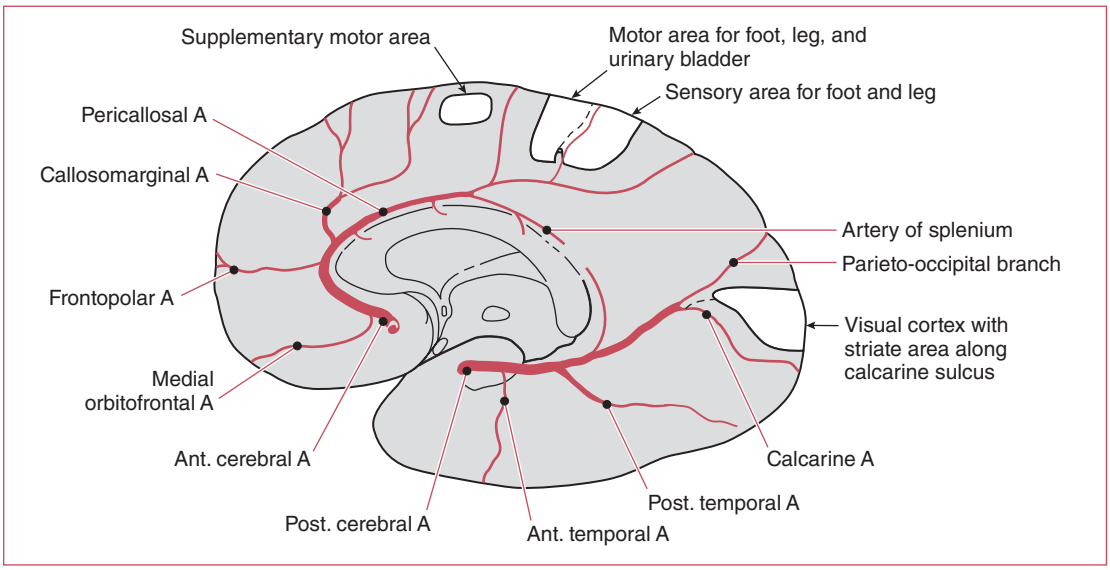


FIGURE 1–8 The distribution of the ACA and PCA on the medial aspect of the cerebral hemisphere, showing principal regions of cerebral localization.
A, artery; ACA, anterior cerebral artery; Ant., anterior; PCA, posterior cerebral artery; Post., posterior.

  - 70% of the time: Both PCAs arise from the basilar artery and are connected to the internal carotids through the posterior communicating artery.
  - 20% to 25%: One PCA comes from basilar, and one PCA comes from ICA.
  - 5% to 10%: Both PCAs arise from carotids.
- Clinical presentation:
  - 🕮 Visual field cuts **(Figure 1–9)**
  - 🕮 Anton syndrome: Visual anosognosia associated with bilateral occipital infarcts, particularly affecting the primary visual cortex and visual association areas. (PCA is the main blood supply.) Cortical blindness with denial of/inability to recognize this loss of vision. May also manifest as homonymous hemianopia, visual neglect, and visual agnosia (De Moraes, 2013; Kwong Yew et al., 2014).
  - May have prosopagnosia (cannot read faces)
  - Palinopsia (abnormal recurring visual imagery)
  - Alexia (cannot read)
  - 🕮 Alexia without agraphia (cannot read or recognize written words but can write) due to injury to the left occipital lobe and splenium of corpus callosum. Left PCA is the artery most commonly involved (Rupareliya et al., 2017).
  - Transcortical sensory aphasia (loss of power to comprehend written or spoken words; patient can repeat)
  - 🕮 Structures supplied by the interpeduncular branches of the PCA include the oculomotor cranial nerve (CN III) and trochlear (CN IV) nuclei and nerves.

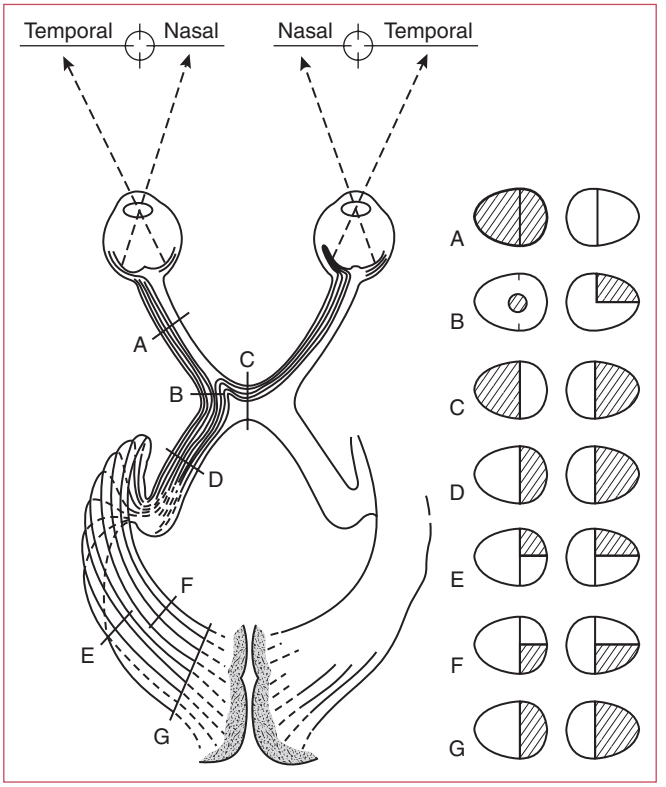


FIGURE 1–9 Diagram showing the effects on the fields of vision produced by lesions at various points along the optic pathway: (A) complete blindness in the left eye from an optic nerve lesion; (B) the usual effect is a left junction scotoma in association with a right upper quadrantanopia. The latter results from interruption of the right retinal nasal fibers that project into the base of the left optic nerve (Wilbrand's knee). A left nasal hemianopia could occur from a lesion at this point but is exceedingly rare; (C) chiasmatic lesion causing bitemporal hemianopia; (D) right homonymous hemianopia from optic tract lesion; (E and F) right superior and inferior quadrant hemianopia from interruption of visual radiations; and (G) right homonymous hemianopia due to lesion of striate cortex.
Source: From Ropper AH, Samuels MA, Klein JP. *Adams and Victor's Principles of Neurology*. 10th ed. McGraw-Hill; 2014, with permission.

– 🕮 Clinical syndromes caused by the occlusion of these branches include *Weber syndrome* (oculomotor palsy with contralateral hemiplegia), which is discussed later, and trochlear nerve palsy (vertical gaze palsy).

VERTEBROBASILAR SYSTEM (FIGURES 1–10 AND 1–11)

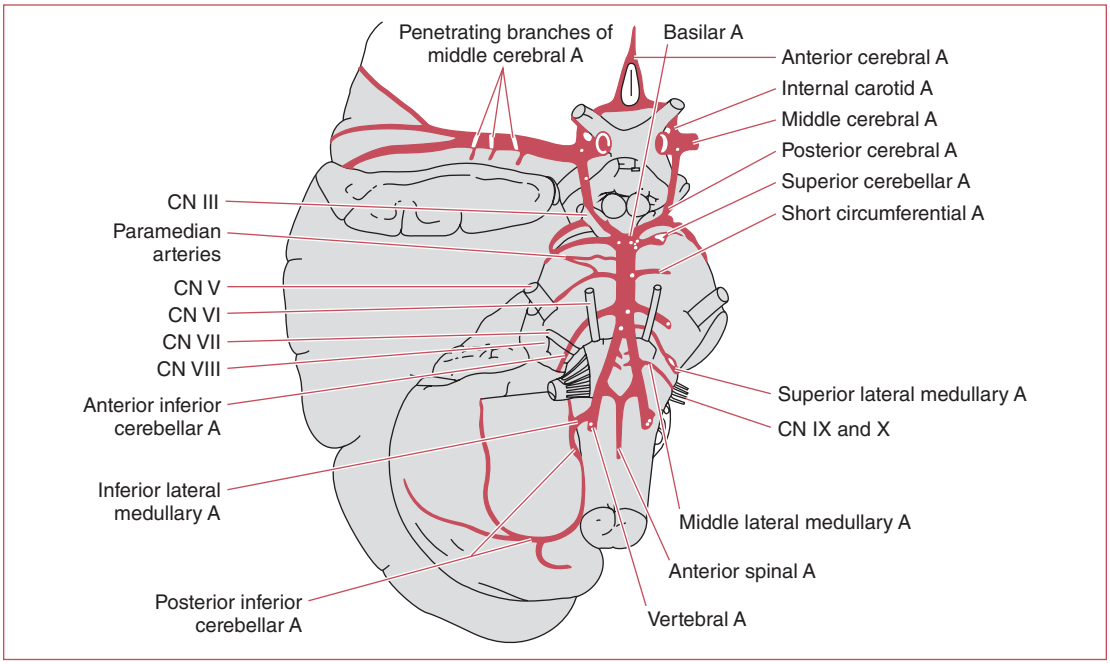


FIGURE 1–10 The principal vessels of the vertebrobasilar system in relation to the brainstem.
A, artery; CN, cranial nerve.

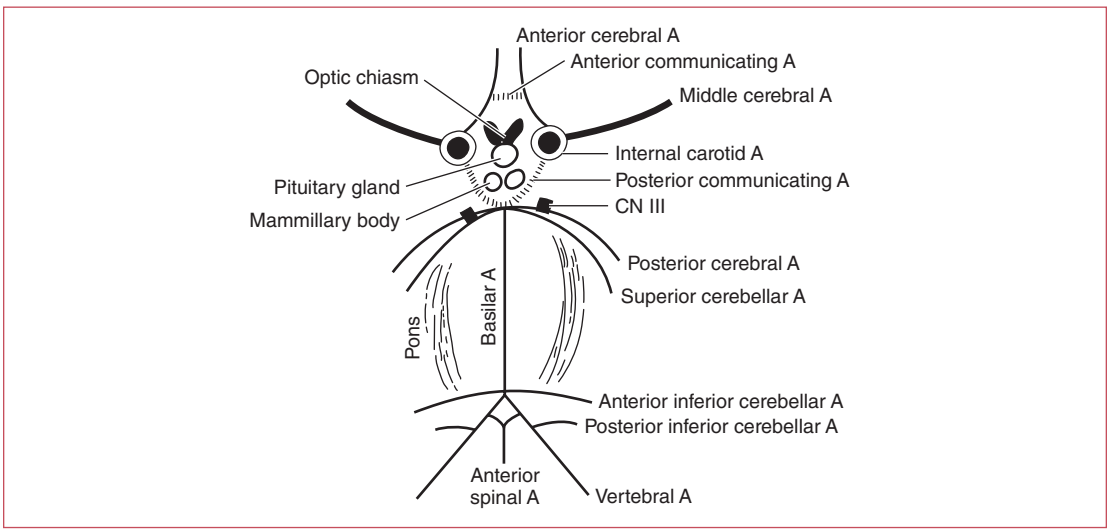


FIGURE 1–11 The Circle of Willis is a ferocious spider that lives in the brain. His name is Willis! Note that he has a nose; angry eyebrows; two suckers; eyes that look outward; a crew cut; antennae; a fuzzy beard; eight legs; a belly that, according to your point of view, is either thin (basilar artery) or fat (the pons, which lies from one end of the basilar artery to the other); two feelers on his rear legs; and male genitalia. Here, the brain is seen from below, so the carotid arteries are seen in cross-section.
A, artery; CN, cranial nerve.
Source: From Goldberg S. *Clinical Neuroanatomy Made Ridiculously Simple*. 5th ed. Medmaster Inc.; 2014.

- The vertebrobasilar arteries supply the midbrain, pons, medulla, cerebellum, and posterior and ventral aspects of the cerebral hemispheres (via the PCAs).
  - Vertebral arteries originate from the subclavian arteries and are the main arteries of the medulla.
  - At the pontomedullary junction, the two vertebral arteries join to form the basilar artery, which provides the blood supply to the pons and midbrain.
  - The cerebellum is supplied by the posterior–inferior cerebellar arteries (PICA) originating from the vertebral arteries as well as the anterior–inferior cerebellar arteries (AICA) and superior cerebellar arteries, both of which originate from the basilar artery.
- Vertebrobasilar system involvement may present with any combination of the following signs/symptoms:
  - Vertigo
  - Nystagmus
  - Abnormalities of motor function, often bilaterally
  - Ipsilateral CN dysfunction
  - 🕮 Crossed signs: Motor or sensory deficit on ipsilateral side of face and contralateral side of body; ataxia, dysphagia, and dysarthria
- 🕮 **Important**: There is an *absence of cortical signs* (such as aphasias or cognitive deficits) in vertebrobasilar system events that would be characteristic of anterior circulation involvement.
- 🕮 **Important**: While isolated attacks of vertigo can be the initial and only symptom of vertebrobasilar insufficiency, attacks of vertigo in vertebrobasilar insufficiency usually last <30 minutes and have no associated hearing loss. If no other symptoms, especially nystagmus, accompany vertigo, a diagnosis of benign paroxysmal positional vertigo (BPPV) should be considered (Bhattacharyya et al., 2008).

SYNDROMES OF THE VERTEBROBASILAR SYSTEM

I. 🕮 Wallenberg (Lateral Medullary) Syndrome

- Wallenberg syndrome is also known as lateral medullary syndrome, PICA syndrome, or vertebral artery syndrome.
- This syndrome is one of the most striking in neurology. It occurs due to occlusion of the following:
  1. Vertebral arteries (involved in eight out of 10 cases)
  2. PICA
  3. Superior lateral medullary artery
  4. Middle lateral medullary artery
  5. Inferior lateral medullary artery
- *Signs and symptoms* include the following (**Table 1–2A**):
  - Ipsilateral side
    - Horner syndrome (ptosis, anhidrosis, and miosis)
    - Decrease in pain and temperature sensation on the ipsilateral face
    - Cerebellar signs such as ataxia on ipsilateral extremities (patient falls to side of lesion)
  - Contralateral side
    - Decreased pain and temperature on contralateral body
    - Dysphagia, dysarthria, hoarseness, and vocal cord paralysis
    - Vertigo, nausea, and vomiting
    - Hiccups
    - Nystagmus and diplopia

Note: No facial or extremity muscle weakness seen in this syndrome.

II. Benedikt Syndrome (Red Nucleus/Tegmentum of Midbrain)

- Obstruction of interpeduncular branches of basilar artery or PCA, or both
- Ipsilateral CN III nerve paralysis with mydriasis, contralateral hypesthesia (medial lemniscus)
- Contralateral hyperkinesia (ataxia, tremor, chorea, and athetosis) due to damage to red nucleus

TABLE 1–2A Symptoms and Site of Lesion of Wallenberg Syndrome

SYMPTOMS	SITE OF LESION
Ipsilateral Horner syndrome (ptosis, miosis, and anhidrosis)	Pupillodilator (sympathetic) fibers
Ipsilateral face pain and temperature loss	Spinal trigeminal tract
Ipsilateral ataxia (UE > LE)	Spinocerebellar tract
Contralateral arm/leg pain and temperature loss	Spinothalamic tract
Dysphagia, hiccups, hoarseness	Nucleus ambiguus, CN IX and CN X nerves
Vertigo, nausea, vomiting	Vestibular nuclei

CN, cranial nerve; LE, lower extremity; UE, upper extremity.
Source: Adapted from Zorowitz RD, Harvey RL. Stroke syndromes. In: Cifu DX, ed. *Braddom's Physical Medicine and Rehabilitation*. 5th ed. Elsevier; 2016:1015.

III. Syndromes of the Paramedian (Medial) Brainstem (Table 1–2B)

- The paramedian area of the brainstem contains:
  - Motor nuclei of CNs
  - Corticospinal tract
  - Medial lemniscus
  - Corticobulbar tract
- Signs/symptoms include:
  - Ipsilateral CN paralysis
  - Contralateral hemiparalysis

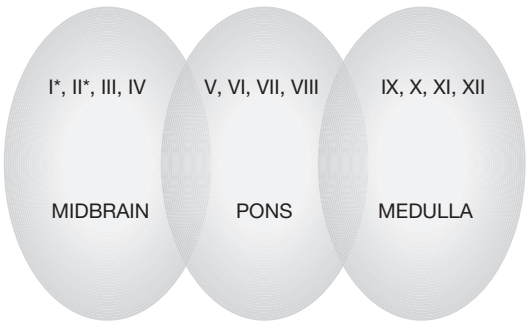
TABLE 1–2B Syndromes of the Paramedian (Medial) Brainstem

SYNDROME	WEBER SYNDROME	MILLARD–GUBLER SYNDROME	MEDIAL MEDULLARY SYNDROME ("ANOTHER LESION")
CN palsy	Ipsilateral CN III palsy	Ipsilateral CN VI paralysis (often CN VII also involved)	Ipsilateral CN XII palsy
Motor deficit	Contralateral hemiplegia	Contralateral hemiplegia (extension into medial lemniscus is Raymond–Foville syndrome with gaze palsy to side of lesion)	Contralateral hemiplegia
Sensory deficit			Contralateral lemniscal sensory loss

CN, cranial nerve.

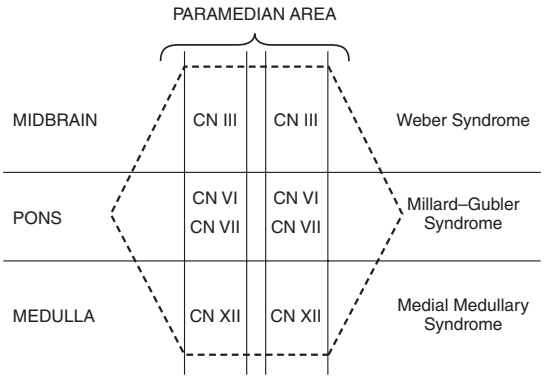
Gross Locations of Cranial Nerve (CN) Brainstem Nuclei

📖 *NOTE: *CN I and *CN II nuclei are located in forebrain. CN XI has two divisions, the cranial and the spinal division. The spinal division of CN XI arises from the ventral horns of C1 through C6 levels.*



Weber Syndrome (Base of Midbrain; Tables 1–2B and 1–3)

- Obstruction of interpeduncular branches of PCA or posterior choroidal artery, or both
- Ipsilateral CN III paralysis
- Contralateral hemiplegia, contralateral Parkinson signs, and contralateral dystaxia (mild degree of ataxia)



GROSS DEPICTION OF THE PARAMEDIAN BRAINSTEM AND ASSOCIATED SYNDROMES

CN, cranial nerve.

Millard–Gubler Syndrome (Base of Pons; Tables 1–2B and 1–3)

- Obstruction of circumferential branches of basilar artery
- Ipsilateral abducens (CN VI) and facial (CN VII) palsies
- Contralateral hemiplegia
- Raymond–Foville syndrome: Lesion of inferior medial pons with clinical signs of ipsilateral CN VI and CN VII palsies; ipsilateral paramedian pontine reticular formation (PPRF) and medial longitudinal fasciculus (MLF) affected leading to conjugate gaze palsy (inability to gaze toward lesion side); also contralateral hemiplegia (lesion to ipsilateral corticospinal tract) and hemisensory loss (lesion to ipsilateral medial lemniscus; Khazaal et al., 2023).

🕮 Medial Medullary Syndrome (Medial Medulla; Tables 1–2B and 1–3)

- Caused by an infarction of the medial medulla due to occlusion (usually atherothrombotic) of the penetrating branches of the vertebral arteries (upper medulla) or anterior spinal artery (lower medulla and medullo-cervical junction)
- Rare; ratio of medial medullary infarct to lateral medullary infarct approximately 1 to 2:10
- Signs and symptoms:
  - Ipsilateral hypoglossal palsy (with deviation toward the side of the lesion)
  - Contralateral hemiparesis
  - Contralateral lemniscal sensory loss (proprioception and position sense)

TABLE 1–3 Brainstem Syndromes

REGION	MAIN ARTERY	MEDIAL BRAINSTEM LESIONS (PARAMEDIAN AREA SYNDROME)	LATERAL BRAINSTEM LESIONS
Midbrain	PCA	Weber syndrome	
Pons	Basilar artery	Millard–Gubler syndrome	
Medulla	Vertebral artery (or anterior spinal artery)	Medial medullary syndrome (“another lesion”)	Wallenberg syndrome

PCA, posterior cerebral artery.

IV. Basilar Artery Occlusion Syndrome

- Occlusion may arise in several ways:
  - Atherosclerotic plaque in the basilar artery itself (usually lower third)
  - Occlusion of both vertebral arteries
  - Occlusion of one vertebral artery when it is the only artery of adequate size

- Note:
  - Thrombosis usually only obstructs a branch of the basilar artery rather than the trunk.
  - If emboli get through the vertebral arteries, they usually lodge in one of the PCAs or at the upper bifurcation of the basilar artery.
  - May cause internuclear ophthalmoplegia, conjugate horizontal gaze palsy, or ocular bobbing. Ptosis and nystagmus are common but variable. May see palatal myoclonus or coma.
- 🕮 ***Locked-in syndrome:*** Tetraparesis with patients who are only able to move their eyes vertically or blink. The patient remains fully conscious secondary to sparing of the reticular activating system (RAS). It is caused by bilateral lesions of the ventral pons (basilar artery occlusion). Some degree of paresis accompanies nearly all cases of basilar artery occlusion. (Note: Majority of RAS is located primarily in the midbrain.)

TABLE 1–4 Neuroanatomic Locations of Lacunar Infarction Syndromes

LACUNAR SYNDROME	ANATOMIC LOCATION
1. Pure motor hemiplegia–Weakness involving face, arm, and leg; no sensory deficits, aphasia, or parietal signs	Posterior limb of internal capsule (supplied by the lenticular striate artery) Corona radiata Pons
2. Pure sensory stroke	Thalamus (ventrolateral) Parietal white matter Thalamocortical projections
3. 🕮 Dysarthria/"clumsy hand syndrome"	Basis pontis Internal capsule (anterior limb)
4. Sensorimotor stroke	Junction of internal capsule and thalamus
5. Ataxia and leg paralysis	Pons Midbrain Internal capsule Cerebellum Parietal white matter Coronal radiata
6. Hemichorea-hemiballismus	Head of the caudate Thalamus Subthalamic nucleus

HEMORRHAGIC STROKES (TABLE 1–1)

- 13% of all strokes may be secondary to HTN, ruptured aneurysm, arteriovenous malformation (AVM), blood dyscrasias/bleeding disorders, anticoagulants, bleeding into tumors, or angiopathies.

I. Hypertensive Intracerebral Hemorrhage

- Linked to chronic HTN (>1/3 occur in normotensives)
- Preceded by formation of "false" aneurysms (microaneurysms) of Charcot and Bouchard = arterial wall dilations secondary to HTN
- Frequently extends to ventricular subarachnoid space
- **Symptoms:**
  - Sudden onset of HA and/or LOC
  - Vomiting at onset in 22% to 44%
  - Seizures occur in 10% of cases (first few days after onset).
  - Nuchal rigidity is common.
- **Locations** include the putamen, thalamus, pons, cerebellum, and cerebrum.
  1. 🕮 **Putamen:** Most common. Hemiplegia secondary to compression of adjacent internal capsule. Vomiting occurs in approximately 50% of patients. HA is frequent but not constant.

- Large hemorrhage: Stupor/coma and hemiplegia with deterioration in hours.
- With smaller hemorrhages: HA leading to aphasia, hemiplegia, and eyes deviate away from paretic limbs.
- These symptoms, occurring over a few minutes to one-half hour, are strongly suggestive of progressive intracerebral bleeding.

2. **Thalamus:** Hemiplegia by compression of adjacent internal capsule; contralateral sensory deficits; aphasia present with lesions of the dominant side; contralateral hemineglect with involvement on the nondominant side. Ocular disturbances with extension of hemorrhage into subthalamus.
3. **Pons:** Deep coma results in a few minutes; total paralysis, small pupils (1 mm) that react to light; decerebrate rigidity → death occurs in a few hours. Patient may survive if hemorrhage is small (<1 cm).
4. **Cerebellum:** Develops over several hours. Coma/LOC; unusual vomiting; occipital HA; vertigo; inability to sit, stand, or walk; eyes deviate to opposite side (ipsilateral CN VI palsy); dysarthria; dysphagia.
5. **Lobar (cerebral) hemorrhage:** HA and vomiting. A study of 26 patients revealed:
   - 11 occipital: Dense homonymous hemianopsia and pain in ipsilateral eye
   - 7 left temporal: Partial hemianopsia/fluent aphasia/ear pain
   - 4 frontal: Contralateral hemiplegia (mainly the arm) and frontal HA
   - 3 parietal: Hemisensory deficit (contralateral)/anterior temporal HA
   - 1 right temporal (Ropper & Davis, 1980)

II. Subarachnoid Hemorrhage (Tables 1–1 and 1–5)

- Typically due to a ruptured saccular arterial aneurysm.
  - Saccular aneurysms = berry aneurysms
- Arterial dilations found at bifurcations of larger arteries at the base of the brain (Circle of Willis or major branches; see **Figure 1–12**).
  - 📖 90% to 95% of saccular aneurysms occur on the anterior part of the Circle of Willis.
  - Presumed to result from congenital medial and elastica defects versus hemodynamic forces, causing focal destruction of internal elastic membrane at bifurcations (Ropper & Samuels, 2009).

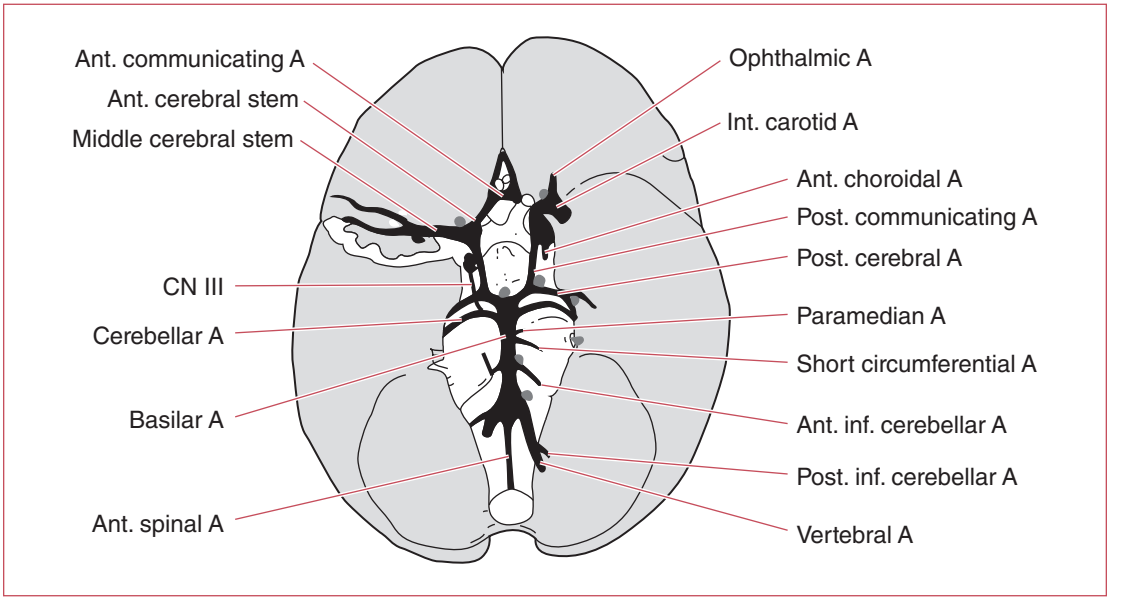


FIGURE 1–12 The principal sites of saccular aneurysms. Gray protrusions at bifurcations of vessels are aneurysms. The majority (approximately 90%) of aneurysms occur in the anterior half of the Circle of Willis.
A, artery; Ant., anterior; CN, cranial nerve; Inf., inferior; Int., internal; Post., posterior.

- Multiple in 20% of patients (either unilateral or bilateral).
- Other types of aneurysms: Arteriosclerotic, mycotic, dissecting aneurysms, traumatic, and neoplastic.
- Risk factors for rupture include HTN, alcohol abuse, drug abuse (e.g., cocaine), smoking, as well as the condition and size of the aneurysm.
- 🕮 More likely to rupture if 10 mm or larger (rupture may occur in smaller aneurysms).
- Rupture occurs usually when patient is active rather than during sleep (e.g., straining, coitus).
- 🕮 Peak age for rupture: Fifth and sixth decade.

TABLE 1–5 Hunt and Hess Scale for Nontraumatic Subarachnoid Hemorrhage Patients

DESCRIPTION	GRADE
Asymptomatic, mild HA, slight nuchal rigidity	1
Moderate to severe HA, nuchal rigidity, no neurologic deficit other than cranial nerve palsy	2
Drowsiness/confusion, mild focal neurologic deficit	3
Stupor, moderate-severe hemiparesis	4
Coma, decerebrate posturing	5

HA, headache.
Source: From Hunt, WE, Hess, RM, Hunt WE, et al., Surgical risk as related to time of intervention in the repair of intracranial aneurysms. *J Neurosurg*. 1968;28(1):14–20.

CLINICAL PRESENTATIONS OF SACCULAR ANEURYSMS/SUBARACHNOID HEMORRHAGE

- Saccular aneurysms are usually asymptomatic prior to rupture. Intracranial aneurysms are common—found in 3% to 5% of patients on routine autopsies.
- 🕮 Compression of adjacent structures such as the oculomotor nerve (CN III) with posterior communicating–internal carotid junction aneurysm or posterior communicating–PCA aneurysm. *Signs of CN III involvement*:
  - Deviation of ipsilateral eye to lateral side (lateral or divergent strabismus) because of unopposed action of lateral rectus muscle
  - Ptosis
  - Mydriasis (dilated pupil) and paralysis of accommodation due to interruption of parasympathetic fibers in CN III.
- Rupture of aneurysm producing SAH with or without intracerebral hematoma.
  - 🕮 "Sentinel" HA: Sudden, intense, and persistent, preceding SAH by days or weeks in approximately 50% of patients.
  - With an SAH, blood is irritating, causing severe HA that is classically described as "worst HA of my life."
  - Sudden, transient LOC in 20% to 45% at onset.
  - Focal neurologic deficits include CN III or CN VI palsy (from direct pressure from the aneurysm vs. accumulation of an intracerebral hematoma vs. early development of arterial spasm), hemiparesis, aphasia (dominant hemisphere), and memory loss.
  - Seizures: 4% at onset/25% overall.
  - 🕮 Mortality: 25% during the first 24 hours.
  - 🕮 Risk of rebleeding within 1 month 30%; 2.2% per year during the first decade.
  - Mortality from rebleeding in the early weeks after initial event: 50% to 60%.
  - Vasospasm: Common complication occurring in approximately 25% of cases; caused by the presence of blood breakdown products (vasoactive amines) on the subarachnoid space acting in the adventitia of the arteries. Occurs 3 to 12 days after rupture (frequently approximately 7 days after rupture).
  - Medications: Nimodipine (calcium channel blocker) is useful in the treatment of cerebral blood vessel spasm after SAH (see Treatment of Subarachnoid Hemorrhage section).

III. Arteriovenous Malformations

- An AVM in the brain consists of a tangled mass of dilated vessels that forms an abnormal communication between the arterial and venous systems.
- These congenital lesions originate early in fetal life.
- AVMs displace rather than invade normal brain tissue.
- 🕮 AVMs are usually low-pressure systems: The larger the shunt, the lower the interior pressure. Thus, with these large dilated vessels, there needs to be an occlusion distally to raise luminal pressures to cause hemorrhage.
- Hemorrhage appears to be more common in smaller malformations, likely due to higher resistance and pressure within these lesions.
- Patients are believed to have a 40% to 50% lifetime risk of hemorrhage from AVM.
- Natural history of AVMs: Risk of bleed approximately 4% per year
- Rebleeding rate 6% to 8% during the first year posthemorrhage
- Annual mortality rate: 1% per year
- First hemorrhage is fatal in approximately 10% of these patients.
- Bleeding commonly occurs between the ages of 20 and 40 years.
- 🕮 **Clinical presentation of AVM rupture**:
  - Hemorrhage is the initial presentation in approximately 50% of cases.
  - May be parenchymal (41%), subarachnoid (23%), or intraventricular (17%) hemorrhage (Brown et al., 1996)
  - Seizures: 30% of cases
  - HAs: 20% of cases; 10% overall with migraine-like HA
  - Neurologic deficit (symptoms vary according to area that is affected)

DIAGNOSTIC STUDIES (TABLE 1–6)

1. Head CT Scan

Major role in evaluating presence of blood (cerebral hemorrhage or hemorrhagic infarction), especially when thrombolysis is being considered.

- If an intracranial hemorrhage is suspected, a head CT *without contrast* is the study of choice.
  - This avoids confusing blood with contrast, as both appear white on CT scan.

TABLE 1–6 Diagnostic Studies

STUDY	ACUTE INFARCT	HEMORRHAGE
CT	**DARK (BLACK)** • Hypodense lesion (dark) = darker than normal • Acute infarct not seen immediately on CT (unless there is a mass effect or large infarct); may be seen after 24 hours (due to increased edema); seen best 3–4 days postinfarct • CT is less sensitive than MRI in detecting acute infarct	**BRIGHT (WHITE)** • Blood seen as hyperdense lesion/ radio-opaque • Seen immediately
MRI	**BRIGHT (WHITE)** • Edema/fluid → hyperintense signal (bright) on T2 • Remember T2 = edema = water (H_2O) • MRI is more sensitive and specific than CT in detecting ischemic infarct	**DARK (BLACK)** • Hypointense on T2 from 0–14 days and >21 days • Other time points (10–21 days), the MRI T2 signal intensity can change to bright or hyperintense • MRI signal changes depending on the acuity/chronicity of the hemorrhage

Ischemic Infarction:

- Regardless of stroke location or size, head CT studies are often normal during the first few hours after a nonhemorrhagic brain infarction.
- The infarcted area appears as a hypodense (dark) lesion usually 24 to 48 hours after the stroke (occasionally positive scans at 3–6 hours → subtle CT changes may be seen early with large infarcts, such as obscuration of gray–white matter junction, sulcal effacement, or early hypodensity).
- Hypodensity is initially mild and poorly defined; edema better seen on the third or fourth day as a well-defined hypodense area.
- Head CT with contrast: Intravenous (IV) contrast provides no brain enhancement on day 1 or 2, as it must wait for enough damage to the blood–brain barrier; more evident in 1 to 2 weeks. Changes disappear 2 to 3 months later.
- Some studies suggest worse prognosis for patients receiving IV contrast early.
- Hemorrhage can occur within an infarcted area, where it will appear as a hyperdense mass within the hypodense edema of the infarct.

Hemorrhagic Infarct/Intracerebral Hemorrhage:

- High-density (white) lesion seen immediately on CT in approximately 100% of cases. Proven to be totally reliable in hemorrhages 1 cm or larger in diameter. Demonstration of clot rupture into the ventricular system (32% in one series) not as ominous as once thought.

Subarachnoid Hemorrhage:

- Positive scan in 90% when CT performed within 4 to 5 days (may be demonstrated for only 8–10 days). SAH can really be visualized only in the acute stage, when blood is denser (whiter) than the CSF.
- Appears as a hyperdense (or isodense) area on CT scan—look for blood in the basal cisterns or increased density in the region around the brainstem. May sometimes localize aneurysm based upon hematoma or uneven distribution of blood in cisterns.
- Once diagnosis of SAH has been established, angiography is generally performed to localize and define the anatomic details of the aneurysm and determine if other aneurysms exist.

2. Brain MRI

- More sensitive than CT scan in detecting acute ischemic infarcts (including small lacunes) and posterior cranial fossa infarcts (images are not degraded by bone artifacts).
  - Edema due to ischemia detected earlier on MRI than with CT—within a few hours of onset of infarct.
  - Diffusion-weighted imaging (DWI) MRI has emerged as the most sensitive and specific imaging technique for acute infarct, far better than CT or any other type of MRI sequence.
  - RAPID (rapid processing of perfusion and diffusion) MRI analyzes and integrates multimode image data mainly based on perfusion-weighted imaging or CT perfusion images, performs well in identifying ischemic penumbra and has been used in many ischemic stroke clinical studies, and promotes transition from "time window" to "tissue window" in the treatment of early-stage acute ischemic stroke (Zhang et al., 2022).

Ischemic Cerebral Infarction:

- DWI has high sensitivity and specificity for detecting infarcted regions, even within minutes of symptom onset.
- Early, hyperintense signal (bright/white) on T2-weighted images, more pronounced at 24 hours to 7 days. T1-weighted images would demonstrate hypointense signal (darker/black) in the same areas.
- Chronic (21 days or more)—decreased on T1 and T2 images.

Intracerebral Hemorrhage

- Acute hemorrhage: Hypointense signal (darker/black) or isointense on T1- and T2-weighted images.
- Edema surrounding hemorrhage appears as hyperintense (brighter/white) on T2 imaging and hypointense (darker) on T1-weighted images.

Subarachnoid or Intraventricular Hemorrhage:
- Acutely, hypointense signal (darker/black) on T1 and T2 images.

Lacunar Infarct:
- CT scan documents most supratentorial lacunar infarctions, while MRI successfully documents both supratentorial and infratentorial infarctions when lacunes are 0.5 cm or greater.

3. Carotid Ultrasound

- Real-time B-mode imaging; direct Doppler examination. Screening test for carotid stenosis; identification of ulcerative plaques less certain. Useful in following patients for progression of stenosis.

4. Transcranial Doppler Ultrasound

- Low-frequency Doppler ultrasound to evaluate basal cranial vessels through temporal bone, orbit, or foramen magnum.
- Velocity and direction of blood flow in all vessels of Circle of Willis may be identified.
- Detects vasospasm and intracranial collateral pathways.

5. Angiography

- Includes conventional angiography, magnetic resonance angiography (MRA), and intra-arterial digital subtraction angiography (DSA). These studies evaluate extracranial and intracranial circulation.
- Valuable tools for diagnosing aneurysms, vascular malformations, arterial dissections, narrowed or occluded vessels, and angiitis.
- Complications occur in 2% to 12%.
  - Include aortic or carotid artery dissection, embolic stroke, vascular spasm, and vascular occlusion
- 2.5% Morbidity associated with the procedure.
- Carotid and vertebral angiography are the only certain means of demonstrating an aneurysm.
  - Positive in 85% of patients with "clinical" SAH
- DSA studies safer; may be performed as outpatient.

MRA:
- Can reliably detect extracranial carotid artery stenosis
- May be useful in evaluating patency of large cervical and basal vessels
- Detects most aneurysms on the basal vessels; insufficient sensitivity to replace conventional angiography

6. Transthoracic Echocardiography and Transesophageal Echocardiography

- Transthoracic echocardiography (TTE) can quickly assess heart valves and ejection fraction.
- Transesophageal echocardiography (TEE) is superior for evaluating aorta, pulmonary artery, heart valves, atria, atrial septum, left atrial appendage, and coronary arteries; TEE also used for detection of PFO.
- Using cardiac catheterization and/or operation as a gold standard, contrast TEE was found to be more sensitive (100% vs. 63%, $p < .005$) and accurate (97% vs. 78%, $p < .05$) than contrast TTE for detection of PFO (Chen et al., 1992).

7. Lumbar Puncture

- Can detect blood in the CSF
- Primarily used in diagnosing a suspected SAH when head CT is not available, or occasionally when the CT is negative and there is still high clinical suspicion.

MEDICAL TREATMENT OF STROKE

IMMEDIATE MANAGEMENT (Ferri, 2010; Rosen, 1992; Stewart, 1999)

- ABCs of critical care: Airway, Breathing, Circulation.
  - **Airway obstruction** can occur with paralysis of the throat, tongue, or mouth muscles and pooling of saliva. Stroke patients with recurrent seizures are at increased risk of airway obstruction. Aspiration is also a concern in hemorrhagic strokes (increased association of vomiting at onset).
  - **Respiratory support:** Breathing abnormalities (central apnea) occasionally seen in patients with severe strokes.
  - **Control of BP**: See following section.
- **IV fluids**: Normal saline solution (NSS) or Ringer's lactate. Avoid hypotonic solutions or excessive fluid loading, as it may worsen brain edema.
- **Keep patient NPO (nothing by mouth)** to avoid risk of aspiration.
- **Emergent head CT scan:**
  - Because the clinical picture of hemorrhagic and ischemic stroke may overlap, head CT scan without contrast is needed in most cases to definitively differentiate between the two.
  - One of the criteria that determines whether a patient is a candidate for emergent thrombolytic therapy.
  - Impaired level of consciousness/coma: If there is acute deterioration of LOC, evaluate for hematoma/acute hydrocephalus. Treatment would be emergency surgery.
  - Rule out hemorrhage in the presence of an underlying coagulopathy.
  - Fever and concern regarding meningitis
- **Seizure management**: See following section.
- **Check blood sugar levels:**
  - Hypoglycemia → bolus 50% IV dextrose
  - Hyperglycemia: Shown to potentiate the severity of brain ischemia in animal studies
  - Insulin if blood sugar >300 mg/dL
- **Check baseline ECG and troponin:** Should not delay administration of tissue plasminogen activator (tPA; Benjamin et al., 2018)
- **Control of intracranial pressure (ICP):** See following section.
- **Fever**: Potentially damaging to the ischemic brain
  - Antipyretics (e.g., acetaminophen) should be given early while the source of fever is being ascertained.

BLOOD PRESSURE MANAGEMENT

- The management of BP after acute ischemic and hemorrhagic stroke is challenging and depends on whether or not the patient is a candidate for thrombolysis. Data to guide recommendations for treatment are inconclusive or conflicting. Many patients have spontaneous declines in BP during the first 24 hours after onset of stroke (Jauch et al., 2013).
- Antihypertensive treatment can lower cerebral perfusion and lead to worsening of stroke. The response of stroke patients to antihypertensive medications can be exaggerated.
- Current treatment recommendations are based on the type of stroke (ischemic vs. hemorrhagic).

Ischemic Stroke Blood Pressure Management (Table 1–7)

- 🕮 IV labetalol, nicardipine, and clevidipine are favored antihypertensive agents. Hydralazine and enalaprilat may also be considered (Benjamin et al., 2018).
- If BP remains uncontrolled or diastolic BP (DBP) >140 mmHg, consider IV sodium nitroprusside (Benjamin et al., 2018).

TABLE 1–7 AHA Recommendations for Hypertension Management in Ischemic Stroke

Nonthrombolytic candidates: The benefit of initiating or reinitiating treatment of hypertension within the first 48–72 hours is uncertain. It might be reasonable to lower BP by 15% during the first 24 hours after onset of stroke (Benjamin et al., 2018).	Treat if:	SBP >220 DBP >120 or MAP >120
Thrombolytic candidates (before thrombolytic treatment given)	Treat if:	SBP >185 DBP >110

AHA, American Heart Association; BP, blood pressure; DBP, diastolic blood pressure; MAP, mean arterial pressure; SBP, systolic blood pressure.
Source: Adapted from American Heart Association. www.heart.org.

Hemorrhagic Stroke Blood Pressure Management

- Treatment of elevated BP during an acute hemorrhagic stroke is controversial. The usual recommendation is to treat at lower levels of BP than for ischemic strokes because of concerns of rebleeding and extension of bleeding.
- Frequent practice is to treat BP if SBP >180 or DBP >105.
- 🕮 Agent of choice: IV labetalol (does not cause cerebral vasodilation, which could worsen increased ICP).

Seizure Management

- Recurrent seizures: Potentially life-threatening complication of stroke (see also Medical Management Problems in Stroke Rehabilitation section)
- Seizures can substantially worsen elevated ICP.
- Benzodiazepines = first-line agents for treating seizures
  - IV lorazepam or diazepam
- If seizures do not respond to IV benzodiazepines, treat with long-acting anticonvulsants:
  - Phenytoin—18 mg/kg
  - Fosphenytoin—17 mg/kg
  - Phenobarbital—1,000 mg or 20 mg/kg

Intercranial Pressure Management

- Increased ICP reduces cerebral perfusion pressure (CPP).
- CPP is calculated by subtracting ICP from mean arterial pressure (MAP).
  - CPP = MAP – ICP
  - CPP should remain >60 mmHg to ensure cerebral blood flow.
- Fever, hyperglycemia, hyponatremia, and seizures can worsen cerebral edema by increasing ICP.
  - ICP 2 to 5 mmHg is considered normal.
  - ICP ≤15 mmHg is considered harmless.

Keep ICP <20 mmHg.

- **Correction of underlying factors exacerbating elevated ICP**:
  - Hypercarbia
  - Hypoxia
  - Hyperthermia
  - Acidosis
  - Hypotension
  - Hypovolemia
- **Positioning:**
  - Avoid flat, supine position; instead, elevate head of bed to 30°.
  - Avoid head and neck positions compressing the jugular veins.

- **Hyperventilation:**
  - Intubation and hyperventilation: Reduction of $PaCO_2$ through hyperventilation is the most rapid means of lowering ICP. Keep ICP <20 mmHg.
  - Hyperventilation should be used with caution because it reduces brain tissue PO_2 ($PbrO_2$). Hypoxia may lead to ischemia of brain tissue, causing further central nervous system (CNS) damage after the stroke.
  - Optimal $PaCO_2$ is approximately 25 to 30 mmHg.
- **Medications:**
  - Hyperosmolar therapy with mannitol improves ischemic brain swelling through diuresis and intravascular fluid shifts.
  - Furosemide/acetazolamide may also be used.
  - High doses of barbiturates (e.g., thiopental) rapidly lower ICP and suppress electrical brain activity.
- **Hypothermia**
- **Fluid restriction:**
  - Avoid glucose solutions; use normal saline; maintain euvolemia.
  - Replace urinary losses with normal saline in patients receiving mannitol.
- **Surgical therapy:**
  - Neurosurgical decompression for lesions with mass effect causing increased ICP

THROMBOLYTIC THERAPY

Intravenous Thrombolytic Therapy

- Intravenous (IV) tissue plasminogen activator (tPA) is the first U.S. Food and Drug Administration (FDA)-approved treatment for acute ischemic strokes in selected patients.
  - Alteplase: Also known as Activase® or recombinant tPA (rtPA). Most commonly used fibrinolytic for acute ischemic stroke (Baig & Bodle, 2024; NIH, 2023; Reed et al., 2024).
  - Tenecteplase (TNK): Has proven similar safety and efficacy to alteplase for treatment of acute ischemic stroke (Kobeissi et al., 2023).
- In the National Institute of Neurological Disorders and Stroke (NINDS) trial (1995), patients given tPA *within 3 hours of onset* of stroke were 30% more likely to have minimal or no disability at 3 months compared with patients treated with placebo.
  - There was a tenfold increase in hemorrhage (overall) with tPA compared with placebo (6.4% vs. 0.6%) and fatal ICH (3% vs. 0.3%).
  - However, overall mortality was higher in the placebo group than in tPA groups: 17% in tPA group (including hemorrhage cases) versus 21% in placebo group.
- In 2008, European Cooperative Acute Stroke Study (ECASS) III demonstrated that IV recombinant tPA (rtPA) can be given safely to and can improve outcomes for carefully selected patients treated 3 to 4.5 hours after stroke (Hacke et al., 2008).
  - The European Medicines Agency expanded the approval of IV rtPA to the 3- to 4.5-hour window, but the FDA has declined to do so.
  - However, the AHA/American Stroke Association (ASA) has recommended tPA in eligible patients who can be treated 3 to 4.5 hours postonset (see the following).

Inclusion Criteria for Tissue Plasminogen Activator Use

- Age 18 years or older
- Time of symptom onset well-established and is <4.5 hours before treatment would begin
- Patients with measurable neurologic deficits (moderate to severe stroke symptoms)
- Head CT negative for blood
- Informed consent of the patient
- Additional inclusion criteria for use of tPA 3 to 4.5 hours postonset (Benjamin et al., 2018):
  - Less than 80 years of age
  - No history of both diabetes mellitus and prior stroke

  - National Institutes of Health Stroke Scale (NIHSS) score ≤25
  - Not taking any oral anticoagulants
  - No imaging evidence of ischemic injury involving more than one-third of the MCA territory

Exclusion Criteria for Tissue Plasminogen Activator Use

- Minor stroke symptoms/TIA (symptoms rapidly improving)
- Head CT positive for blood
- BP >185/100 despite medical treatment
- Coagulopathy/patient on anticoagulants (warfarin, antiplatelet agents, and heparin)
  - Prothrombin time (PT) >15 seconds or international normalized ratio (INR) >1.7
  - Patient received heparin within 48 hours prior with elevated activated partial thromboplastin time (APTT)
  - Patient taking warfarin
- Platelet count <100,000
- Blood sugar <50 or >400
- History of stroke or severe head injury in the past 3 months
- History of ICH, AVM, or aneurysm
- History of gastrointestinal (GI) or genitourinary (GU) bleeding within the past 21 days
- No pregnancy or lactation within the past 30 days
- Major surgery within the past 14 days
- Seizure at onset of stroke
- Acute MI

Intra-Arterial Thrombolytic Therapy (Benjamin et al., 2018)

- Initial treatment with intra-arterial thrombolysis is beneficial for carefully selected patients with major ischemic strokes of <6 hours' duration caused by occlusions of the MCA, but data are derived from clinical trials that no longer reflect current practice, including the use of fibrinolytic drugs that are not available.
- Intra-arterial thrombolysis initiated within 6 hours of stroke onset in carefully selected patients who have contraindications to the use of IV tPA might be considered, but the consequences are unknown.
- A clinically beneficial dose of intra-arterial tPA is not established, and tPA does not have FDA approval for intra-arterial use.
- Mechanical thrombectomy with stent retrievers is recommended over intra-arterial thrombolysis as first-line therapy.

Intra-Arterial Mechanical Thrombectomy

- Thrombectomy devices can be useful in achieving recanalization alone or in combination with pharmacologic fibrinolysis in carefully selected patients.
- Stent retrievers (e.g., Solitaire™ FR, Trevo®) are generally preferred to coil retrievers (e.g., Merci®).
- The usefulness of mechanical thrombectomy devices other than the Merci® Retriever, the Penumbra® System, Solitaire™ FR, and Trevo® is not well-established.
- In patients who are potential candidates for mechanical thrombectomy, imaging of the extracranial carotid and vertebral arteries, in addition to the intracranial circulation, is reasonable to provide useful information on patient eligibility and endovascular procedural planning (Benjamin et al., 2018).
- In selected patients with acute ischemic stroke within 6 to 24 hours of last known normal who have large-vessel occlusion in the anterior circulation, obtaining CT, DWI-MRI, or MRI perfusion is recommended to aid in patient selection for mechanical thrombectomy, but only when imaging and other eligibility criteria from randomized clinical trials showing benefit are being strictly applied in selecting patients for mechanical thrombectomy (Benjamin et al., 2018).

ANTICOAGULANT THERAPY

Heparin, Low-Molecular-Weight Heparin, Warfarin

- Immediate anticoagulant therapy in patients with acute ischemic stroke is not associated with any significant net short- or long-term benefit.
- Urgent anticoagulation, with the goal of preventing early recurrent stroke, halting neurologic worsening, or improving outcomes after acute ischemic stroke, is not recommended for treatment of patients with acute ischemic stroke (Benjamin et al., 2018).
- The usefulness of urgent anticoagulation in patients with severe stenosis of an ICA ipsilateral to an ischemic stroke is not well-established (Benjamin et al., 2018).
- The safety and usefulness of short-term anticoagulation for nonocclusive, extracranial intraluminal thrombus in the setting of acute ischemic stroke are not well-established (Benjamin et al., 2018).
- In patients with AFib or hypercoagulable states who have had a recent stroke or TIA, long-term oral anticoagulation is preferred.

Indications for Anticoagulation

- 🕮 **Cardiac emboli:** Best reason to anticoagulate.
  - Primarily from nonvalvular AFib and mural thrombus from MI (Ryder & Benjamin, 1999)
    - Among patients with nonvalvular AFib, anticoagulation can reduce stroke by 60% (Hart et al., 2007).
  - Timing of anticoagulation in patients with cardiac emboli is controversial; probable risk of inducing cerebral hemorrhage or hemorrhagic infarction within large infarcts if anticoagulated in the first 24 to 36 hours.
  - If neurologic deficit is mild (and CT shows no hemorrhage), may begin anticoagulation immediately.
  - If deficit severe (clinically and/or with CT), wait 3 to 5 days before starting anticoagulation.
  - 75% of cardiogenic emboli lodge in the brain. The most common cause is chronic AFib.
- **TIA:**
  - Full anticoagulation in TIA is indicated only with cardioembolic sources and in crescendo TIA, while performing a rapid workup.
- **Completed stroke:**
  - Anticoagulation is not considered beneficial after major infarction and usually not of great value once stroke is fully developed.
  - Anticoagulation is generally not employed for lacunar infarction.
- **Anticoagulation agents:**
  - Warfarin (Coumadin®) inhibits vitamin K-dependent coagulation factors.
  - Dabigatran (Pradaxa®) is a direct thrombin inhibitor indicated to reduce the risk of stroke and systemic embolism in patients with nonvalvular AFib (FDA-approved October 2010).
  - Rivaroxaban (Xarelto®) is a factor Xa inhibitor indicated to reduce the risk of stroke and systemic embolism in nonvalvular AFib (FDA-approved November 2011).
  - Apixaban (Eliquis®) is a factor Xa inhibitor anticoagulant indicated to reduce the risk of stroke and systemic embolism in patients with nonvalvular AFib (FDA-approved December 2012).

ANTIPLATELET THERAPY

Aspirin, Clopidogrel (Benjamin et al., 2018)

- Aspirin administration is recommended in patients with acute ischemic stroke within 24 to 48 hours after onset.
- For those treated with tPA, aspirin administration is generally delayed until 24 hours later but might be considered in the presence of concomitant conditions for which such treatment given

in the absence of tPA is known to provide substantial benefit or withholding such treatment is known to cause substantial risk.
- In patients presenting with minor stroke, treatment for 21 days with dual antiplatelet therapy (aspirin and clopidogrel) that is started within 24 hours can be beneficial for early secondary stroke prevention for a period of up to 90 days from symptom onset (Wang et al., 2013).

CORTICOSTEROIDS

- No utility in ischemic strokes.
- Some studies suggest worsening in prognosis of stroke patients due to hyperglycemia.

CAROTID ENDARTERECTOMY (Moore, 1995)

Symptomatic Carotid Stenosis

- 🕮 CEA for symptomatic lesions with >70% stenosis (70%–99%) is effective in reducing the incidence of ipsilateral hemisphere stroke (endarterectomy for moderate symptomatic carotid stenosis: interim results from the Medical Research Council [MRC] European Carotid Surgery Trial [ECS], 1996; North American Symptomatic Carotid Endarterectomy Trial [NASCET] Collaborators, 1991; Rerkasem & Rothwell, 2011; Walker et al., 1995).
- CEA reduced the risk of disabling stroke as well as death for patients with stenosis exceeding 70% in the ECS trial or 50% in the NASCET.
- Results generalizable only to surgically fit patients operated on by surgeons with complication rates <6%.

Asymptomatic Carotid Stenosis (Chambers & Donnan, 2005)

- Despite about a 3% perioperative stroke or death rate, CEA for asymptomatic carotid stenosis reduces the risk of ipsilateral stroke and any stroke by approximately 30% over 3 years.
- However, absolute risk reduction is small (approximately 1% annually over the first few years of follow-up) but could be higher with longer follow-up (Young et al., 1996).

TREATMENT OF SUBARACHNOID HEMORRGAGE (SEE ALSO INTRACRANIAL PRESSURE MANAGEMENT SECTION)

- Bedrest in a quiet, dark room with cardiac monitoring (cardiac arrhythmias are common)
- Control of HAs with acetaminophen and codeine
- Mannitol to reduce cerebral edema
- BP control. Also have the patient avoid all forms of straining (give stool softeners and mild laxatives).
- Early surgery with clipping or coiling of the aneurysm reduces risk of rebleeding but does not prevent vasospasm or cerebral ischemia.
- 🕮 Nimodipine (calcium channel blocker) decreases cerebral vasospasms after SAH and has been shown to improve outcome after SAH. It also decreases the incidence of permanent neurologic damage and death.
  - Therapy should be initiated within 96 hours of onset of hemorrhage.
  - Treatment: Nimodipine (Nimotop®) 60 mg PO every 4 hours × 21 days.

TREATMENT OF INTRACEREBRAL HEMORRHAGE

- Management of increased ICP and BP (see earlier sections in this chapter).
- Large intracranial or cerebellar hematomas often require surgical decompression.
- The 2022 AHA/ASA guidelines recommend ICP monitoring in ICH patients with a Glasgow Coma Scale (GCS) score of 8 or less (Greenberg et al., 2022).

TREATMENT OF ARTERIOVENOUS MALFORMATIONS (Hamilton & Spetzler, 1994; Schaller et al., 1998)

- Treatment advised in both *symptomatic* and *asymptomatic* AVMs
- Surgical excision if size and location feasible (and depending on perioperative risk)
- Embolization
- Proton beam therapy (via stereotaxic procedure)
- Small asymptomatic AVMs: Radiosurgery/microsurgical resection recommended

TREATMENT OF PATENT FORAMEN OVALE

- The RESPECT trial showed that PFO closure was superior to medical management in reducing recurrent ischemic strokes on long-term follow-up (>5 years) in patients with true cryptogenic stroke and evidence of a PFO (Saver et al., 2017).

STROKE REHABILITATION

INTRODUCTION

The primary goal of stroke rehabilitation is functional restoration by maximizing the independence, lifestyle, and dignity of the patient. This approach implies rehabilitative efforts from a physical, behavioral, cognitive, social, vocational, adaptive, and re-educational point of view. The multidimensional nature of stroke and its consequences make coordinated and combined interdisciplinary team care the most appropriate strategy to treat the stroke patient.

Recovery From Impairments

- Hemiparesis and motor recovery have been the most studied of all stroke impairments. Up to 88% of acute stroke patients have some degree of hemiparesis.
- The process of recovery from stroke-induced hemiparesis usually follows a relatively predictable, stereotyped series of events. This sequence of events has been systematically described by several clinical researchers.
- Twitchell (1951) published a highly detailed report describing the pattern of motor recovery following a stroke (Note: Pattern most consistent in patients with cerebral infarction in the MCA distribution.) His sample included 121 patients; all except three having suffered either thrombosis or embolism of one of the cerebral vessels.
  - Immediately following onset of hemiplegia, there is total loss of voluntary movement and loss or decrease of tendon reflexes.
  - Within 48 hours, increased deep tendon reflexes on the involved side develop.
  - This is followed by the return and then increase of muscle tone against passive movement (tone returns → spasticity), especially in the flexors and adductors in the UE and the extensors and adductors in the lower extremity (LE) on the hemiparetic side.
  - As spasticity increases, clonus of the ankle plantar flexors appears in 1 to 38 days postonset of hemiplegia.
  - Recovery of movement:
    - 6 to 33 days after the onset of hemiplegia, the first "intentional" movements (shoulder flexion) appear.
    - In the UE, a flexor synergy pattern develops (with shoulder, elbow, wrist, and finger flexion) followed by the development of an extensor synergy pattern. Voluntary movement in the lower limb also begins with flexor synergy (also proximal hip) followed by extensor synergy pattern.
- With increased voluntary movement, there is a decrease in the spasticity of the muscles involved.

- Tendon reflexes remain increased despite complete recovery of movement.
- At onset of hemiplegia, the arm is more involved than the leg. Eventual motor recovery in the leg occurs earlier and is more complete than in the arm.
- Most recovery takes place in the first 3 months and only minor additional recovery occurs after 6 months postonset.

Predictors of Motor Recovery (Twitchell, 1951)

- Severity of UE weakness at onset
  - With complete arm paralysis at onset, there is poor prognosis of recovery of useful hand function (only 9% gain good recovery of hand function).
- Timing of return of hand movement
  - If the patient shows some motor recovery of the hand by 4 weeks, there is up to a 70% chance of making a full or good recovery.
- Poor prognosis also associated with:
  - No measurable grasp strength by 4 weeks
  - Severe proximal spasticity
  - Prolonged "flaccidity" period
  - Late return of proprioceptive facilitation (tapping) response >9 days
  - Late return of proximal traction response (shoulder flexors/adductors) >13 days
- Brunnstrom (1966) as well as Sawner and LaVigne (1992) also described the process of recovery following stroke-induced hemiplegia. The process was divided into a number of stages.
  1. **Flaccidity** (immediately after the onset)
     No "voluntary" movements on the affected side can be initiated.
  2. **Spasticity appears**
     Basic synergy patterns appear.
     Minimal voluntary movements may be present.
  3. **Patient gains voluntary control over synergies**.
     Increase in spasticity
  4. **Some movement patterns out of synergy are mastered**.
     Synergy patterns still predominate.
     Decrease in spasticity
  5. **If progress continues, more complex movement combinations are learned as the basic synergies lose their dominance over motor acts**.
     Further decrease in spasticity
  6. **Disappearance of spasticity**
     Individual joint movements become possible and coordination approaches normal.
  7. **Normal function is restored.**

🕮 REHABILITATION METHODS FOR MOTOR DEFICITS

Major Theories of Rehabilitation Training

TRADITIONAL THERAPY

A traditional therapeutic exercise program consists of positioning, range of motion (ROM) exercises, strengthening, mobilization, compensatory techniques, and endurance training (e.g., aerobics). Traditional approaches for improving motor control and coordination emphasize the need for repetition of specific movements for learning the importance of sensation to the control of movement, and the need to develop basic movements and postures (Kirsteins et al., 1999).

PROPRIOCEPTIVE NEUROMUSCULAR FACILITATION (Knott & Voss, 1968)

- 🕮 Uses spiral and diagonal components of movement rather than the traditional movements in cardinal planes of motion with the goal of facilitating movement patterns that will have more functional relevance than the traditional technique of strengthening individual group muscles.

- Theory of spiral and diagonal movement patterns arose from observations that the body will use muscle groups synergistically related (e.g., extensors vs. flexors) when performing a maximal physical activity.
- Stimulation of nerve/muscle/sensory receptors to evoke responses through manual stimuli to increase ease of movement-promotion function.
- Resistance is used during the spiral and diagonal movement patterns with the goal of facilitating "irradiation" of impulses to other parts of the body associated with the primary movement (through increased membrane potentials of surrounding alpha motoneurons, rendering them more excitable to additional stimuli and thus affecting the weaker components of a given part).
- Mass movement patterns keep Beevor's axiom: The brain knows nothing of individual muscle action but only movement.

BOBATH APPROACH/NEURODEVELOPMENTAL TECHNIQUE (Bobath, 1978)

- 🕮 The goal of neurodevelopmental technique (NDT) is to normalize tone, inhibit primitive patterns of movement, and facilitate automatic, voluntary reactions as well as subsequent normal movement patterns.
- Probably the most commonly used approach.
- Based on the concept that pathologic movement patterns (limb synergies and primitive reflexes) must not be used for training, because continuous use of the pathologic pathways may make it too readily available to use at the expense of the normal pathways.
- Suppress abnormal muscle patterns before normal patterns are introduced.
- Mass synergies are avoided, although they may strengthen weak, unresponsive muscles, because these reinforce abnormally increased tonic reflexes and spasticity.
- Abnormal patterns are modified at *proximal* key points of control (e.g., shoulder and pelvic girdle).
- *Opposite* to the Brunnstrom approach, which encourages the use of abnormal movements; see the following section.

BRUNNSTROM APPROACH/MOVEMENT THERAPY (Sawner & LaVigne, 1992)

- 🕮 Uses primitive synergistic patterns in training in an attempt to improve motor control through central facilitation.
- Based on the concept that damaged CNS regresses to phylogenetically older patterns of movements (limb synergies and primitive reflexes). Thus, synergies, primitive reflexes, and other abnormal movements are considered normal processes of recovery before normal patterns of movements are attained.
- Patients are taught to use and voluntarily control the motor patterns available to them at a particular point during their recovery process (e.g., limb synergies).
- Enhances specific synergies through use of cutaneous/proprioceptive stimuli, central facilitation using Twitchell's recovery.
- *Opposite* to the Bobath approach, in which the goal is to inhibit abnormal patterns of movement.

SENSORIMOTOR APPROACH/ROOD APPROACH (Schultz-Krohn, 2013)

- 🕮 Modification of muscle tone and voluntary motor activity using cutaneous sensorimotor stimulation.
- Facilitatory or inhibitory inputs through the use of sensorimotor stimuli, including quick stretch, icing, fast brushing, slow stroking, tendon tapping, vibration, and joint compression, to promote contraction of proximal muscles.

MOTOR RELEARNING PROGRAM/CARR AND SHEPHERD APPROACH (Carr et al., 1985)

- Based on cognitive motor relearning theory and influenced by the Bobath approach.
- Goal is for the patient to relearn how to move functionally and how to problem-solve during attempts at new tasks.

- Instead of emphasizing repetitive performance of a specific movement for improving skill, it teaches general strategies for solving motor problems.
- Emphasizes functional training of specific tasks, such as standing and walking, and carryover of those tasks.

COMPARISON OF THEORIES (Pollock et al., 2014)

- No one approach to physical rehabilitation is any more or any less effective in promoting recovery of function and mobility after stroke.
- Evidence indicates that physical rehabilitation should not be limited to compartmentalized, named approaches, but rather should comprise clearly defined, well-described, evidence-based physical treatments, regardless of historical or philosophical origin.

OTHER APPROACHES

- *Constraint-induced movement therapy (CIMT)* has been statistically shown to produce clinically significant improvements in arm motor function that persist >1 year (EXCITE trial; Wolf et al., 2006).
  - CIMT requires that patients be able to extend their wrists and actively move their digits.
  - 🕮 In the EXCITE trial, participants were required to have at least 10° active wrist extension, at least 10° thumb abduction/extension, and at least 10° extension in at least two additional digits.
- *Body-weight-support treadmill training* was not shown to be superior to progressive exercise at home managed by a physical therapist (LEAPS trial; Duncan et al., 2011).
  - Subjects who received body-weight-support treadmill training within 2 months after stroke were at higher risk of falls than those in other groups.
- *Functional electrical stimulation (FES)* may improve the ability to voluntarily move the affected limb and/or use the affected limb in everyday activities (Pomeroy et al., 2006).
  - The available evidence suggests there might be a small effect on some aspects of function in favor of electrical stimulation compared with no treatment.
  - Currently, there are insufficient data to support or refute the clinical use of FES for neuromuscular retraining.
- *Electromyographic biofeedback (EMG-BF)* makes patient aware of muscle activity or lack of it by using external representation (e.g., auditory or visual cues) of internal activity as a way to assist in the modification of voluntary control.
  - In addition to trying to modify autonomic function, EMG-BF also attempts to modify pain and motor disturbances by using volitional control and auditory, visual, and sensory clues.
  - Electrodes are placed over agonists/antagonists for facilitation/inhibition.
  - Accurate sensory information reaches the brain through systems unaffected by brain → via visual and auditory for proprioception.
  - 🕮 EMG-BF may require adequate visual, auditory, communication, and cognitive skills for active participation by the patient and re-education given by the therapist. Proper evaluation of patients with aphasia, cognitive deficits, and sensory impairment including proprioceptive is recommended (Frontera & DeLisa, 2019).
  - EMG-BF is not advised in cases of complete paralysis (Malik & Dua, 2022).
  - Despite evidence from a small number of individual studies suggesting that EMG-BF plus standard PT produces improvements in motor power, functional recovery, and gait quality when compared with standard physiotherapy alone, combination of all the identified studies did not find a treatment benefit. Overall, the results are limited because the trials were small, generally poorly designed, and utilized varying outcome measures (Woodford & Price, 2007).
- Robotic devices are being developed to improve the rehabilitation of extremities by providing passive and active ROM and measurement of improvements in mobility and strength.

  - Examples: AutoAmbulator and treadmill-supported orthosis
  - There is insufficient evidence to support or refute use in stroke rehabilitation.
- *Motor imagery* is a mental process during which an individual rehearses or simulates a given action before it is actually performed.
  - There is limited evidence to suggest that motor imagery in combination with other rehabilitation treatment appears to be beneficial in improving UE function after stroke, as compared with other rehabilitation treatments without motor imagery. Evidence regarding improvement in motor recovery and quality of movement is less clear (Barclay-Goddard et al., 2011).
- Bilateral arm training hypothesizes that there is a coupling effect that reinforces a possible training benefit to the affected limb when bimanual tasks are performed.
  - There is insufficient good-quality evidence to make recommendations about the relative effect of simultaneous bilateral training compared with placebo, no intervention, or usual care (Coupar et al., 2010).
- *Mirror therapy* utilizes a mirror placed in the patient's midsagittal plane, thus reflecting movements of the nonparetic side as if it were the affected side.
  - The results indicate evidence for the effectiveness of mirror therapy in improving UE motor function, activities of daily living (ADLs), and pain, at least as an adjunct to normal rehabilitation for patients after stroke.
  - Limitations are due to small sample sizes of most included studies, control interventions that are not used routinely in stroke rehabilitation, and some methodologic limitations of the studies (Thieme et al., 2012).
- *Virtual reality* utilizes computer-simulated environment and interactive video gaming to provide patients with engaging activities to improve motor or cognitive function.
  - The use of virtual reality and interactive video gaming was not more beneficial than conventional therapy approaches in improving upper limb function.
  - May be beneficial in improving upper limb function and ADL function when used as an adjunct to usual care (to increase overall therapy time).
  - Insufficient evidence to reach conclusions about the effect of virtual reality and interactive video gaming on gait speed, balance, participation, or quality of life (Laver et al., 2017).
- *Noninvasive brain stimulation* includes repetitive transcranial magnetic stimulation (rTMS) and transcranial direct current stimulation (tDCS; Sandrini & Cohen, 2013).
  - Can be used to modulate cortical excitability during and for several minutes after the end of the stimulation period.
  - Cortical excitability can be reduced (inhibition) or enhanced (facilitation) depending upon parameters.
  - Current evidence does not support the routine use of rTMS for the treatment of stroke (Hao et al., 2013).
  - Evidence on the effectiveness of tDCS (anodal/cathodal/dual) versus control (sham/any other intervention) for improving ADL performance after stroke of very low-to-moderate quality (Elsner et al., 2016).
  - No evidence of the effectiveness of tDCS (anodal tDCS, cathodal tDCS, and bihemispheric tDCS) versus control (sham tDCS) for improving functional communication, language impairment, and cognition in people with aphasia after stroke (Elsner et al., 2015).
  - *Vagus nerve stimulation (VNS)* is a form of neuromodulation therapy with an implanted device that directly stimulates the cervical vagus nerve (Xie et al., 2021).
  - Thought to improve motor function by enhancing neuroplasticity through neuromodulatory effects on the release of acetylcholine and norepinephrine in the cerebral cortex in people with chronic arm weakness after ischemic stroke (Keser & Feng, 2023).
  - Dawson et al., 2021:
    - 108 patients (53 in VNS group vs. 55 in control group; both groups underwent surgical implantation of a stimulator but the VNS group subsequently had active vagus nerve stimulation while the control group received sham stimulation.)

    - FMA-UE (Fugl-Meyer Assessment-Upper Extremity) scores: VNS group increased by 5.0 points (SD 4.4); control group increased by 2.4 points (3.8; $p = .0014$).
    - Clinically meaningful response of FMA-UE score 90 days after in-clinic therapy: 23/53 (47%) in the VNS group; 13/55 (24%) in the control group ($p = .0098$)
  - Implantable VNS, when paired with a rehabilitation program, was effective in improving upper limb motor function in patients who had an ischemic stroke (Malakouti et al., 2024).
- *Medications*:
  - Serotonin selective reuptake inhibitors (SSRIs):
    - FLAME trial: Early prescription of fluoxetine with physiotherapy enhanced motor recovery after 3 months (Chollet et al., 2011).
    - SSRIs appeared to improve dependence, disability, neurologic impairment, anxiety, and depression after stroke, but there was heterogeneity between trials and methodologic limitations in a substantial proportion of the trials. Large, well-designed trials are now needed to determine whether SSRIs should be given routinely to patients with stroke (Mead et al., 2012).
    - A systematic review and meta-analysis concluded that SSRIs are effective in preventing and treating depression, and improving anxiety, motor function, cognitive function, and dependence in patients after stroke. These benefits were only reproducible with the citalopram subanalysis but not fluoxetine. Additional trials are needed to evaluate the safety, efficacy, and timing of SSRIs in patients after stroke (Kalbouneh et al., 2022).
  - Amphetamines: Too few patients have been studied to draw any definite conclusions about the effects of amphetamine treatment on recovery from stroke (Martinsson et al., 2007).
- *Stem cell implantation*: Stem cells have been administered through various routes (intravenously or intra-arterially is preferred during the acute phase poststroke, or intracerebrally in the chronic phase). It is suspected the benefit may lie in immunomodulation, reducing acute tissue injury (acute), or stimulating brain remodeling (chronic; Boncoraglio et al., 2019).
  - Stem cell use was associated with a reduced neurologic impairment, but not with better functional outcome. However, more well-designed trials are needed as certainty of the evidence ranged from low to very low and conclusions came mostly from small randomized controlled trials with high risk of bias.

🕮 POSTSTROKE SHOULDER PAIN (TABLE 1–8) (Lombard et al., 2009)

- 70% to 84% of stroke patients with hemiplegia have shoulder pain with varying degrees of severity.
- Of the patients with shoulder pain, the majority (85%) will develop it during the spastic phase of recovery.
- It is generally accepted that the most common causes of hemiplegic shoulder pain are complex regional pain syndrome (CRPS) type I (see CRPS Type I section) and soft tissue lesions (including plexus lesions).

🕮 Complex Regional Pain Syndrome Type I (CRPS Type I) (Also see the CRPS section in Chapter 11, Pain Medicine)

- CRPS refers to neuropathic pain disorders characterized by an exaggerated response to a traumatic lesion or peripheral nerve that results in severe neuropathic pain, as well as sensory, autonomic, motor, and trophic impairments (Harden et al., 2010). This includes sympathetic-maintained pain and related sensory abnormalities, abnormal blood flow, abnormalities in the motor system, and changes in both superficial and deep structures with trophic changes.
- CRPS type I is formerly known as reflex sympathetic dystrophy (RSD), shoulder-hand syndrome, or Sudeck atrophy.

- CRPS type I follows an injury without nerve injury in the affected limb, whereas CRPS type II develops following a peripheral nerve injury to the affected limb.
- Reported in 12% to 25% of hemiplegic stroke patients. The most common subtype of CRPS in stroke is shoulder-hand syndrome.

STAGES OF CRPS

- **Stage 1 (acute):** Burning pain, diffuse swelling/edema, exquisite tenderness, hyperpathia and/or allodynia, vasomotor changes in hand/fingers (increased nail and hair growth, hyperthermia or hypothermia, sweating). Lasts 3 to 6 months.
- **Stage 2 (dystrophic):** Pain becomes more intense and spreads proximally, skin/muscle atrophy, brawny edema, cold insensitivity, brittle nails/nail atrophy, decreased ROM, mottled skin, early atrophy, and osteopenia (late). Lasts 3 to 6 months.
- **Stage 3 (atrophic):** Pain decreases; trophic changes occur; hand/skin appear pale and cyanotic with a smooth, shiny appearance, feeling cool and dry; bone demineralization progresses with muscular weakness/atrophy, contractures/flexion deformities of shoulder/hand, tapering digits; no vasomotor changes.

PATHOGENESIS

- Multiple theories postulated.
  - Abnormal adrenergic sensitivity develops in injured nociceptors, and circulating or locally secreted sympathetic neurotransmitters trigger the painful afferent activity.
  - Cutaneous injury activates nociceptor fibers → central pain-signaling system → pain.
  - Central sensitization of pain-signaling system
  - Low-threshold mechanoreceptor input develops capacity to evoke pain.
  - With time, efferent sympathetic fibers develop capacity to activate nociceptor fibers.

DIAGNOSIS

- 🕮 Diagnostic criteria for CRPS (the "Budapest Criteria") (Harden et al., 2010):
  - "Continuing pain, which is disproportionate to any inciting event
  - Must report at least one symptom in three of four of the following categories:
    - Sensory—reports of hyperaesthesia and/or allodynia
    - Vasomotor—reports of temperature asymmetry and/or skin color changes and/or skin color asymmetry
    - Sudomotor/edema—reports of edema and/or sweating changes and/or sweating asymmetry
    - Motor/trophic—reports of decreased range of motion and/or motor dysfunction (weakness, tremor, and dystonia) and/or trophic changes (hair, nail, and skin)
  - Must display at least one sign at time of evaluation in two or more of the following categories:
    - Sensory—evidence of hyperalgesia (to pinprick) and/or allodynia (to light touch and/or deep somatic pressure and/or joint movement)
    - Vasomotor—evidence of temperature asymmetry and/or skin color changes and/or asymmetry
    - Sudomotor/edema—evidence of edema and/or sweating changes and/or sweating asymmetry
    - Motor/trophic—evidence of decreased range of motion and/or motor dysfunction (weakness, tremor, and dystonia) and/or trophic changes (hair, nail, and skin)
  - There is no other diagnosis that better explains the signs and symptoms."
- **X-rays**—normal in initial stages; periarticular osteopenia may be seen in later stages. Use is questionable given that bone mineral density starts to decrease in the paralytic arm 1 month after stroke.
  - Need 30% to 50% demineralization for detection

- **Triple-phase bone scan**—30 stroke survivors <3 months from onset were evaluated for CRPS type I using triple-phase bone scan (Harbert et al., 1996; Kozin et al., 1981; Simon & Carlson, 1980).
  - Sensitivity approximately 92%
  - Specificity approximately 56%
  - Positive predictive value (PPV) approximately 58%
  - Negative predictive value (NPV) approximately 91% (Holder & Mackinnon, 1984)
  - Diffusely increased juxta-articular tracer activity on delayed images is the most sensitive indicator for RSD (sensitivity 96%, specificity 97%, and PPV 88%).
- **EMG—**as predictor for CRPS (Cheng & Hong, 1995)
  - Association between spontaneous activity and eventual development of CRPS (vs. no spontaneous activity on EMG)
- **Clinical exam** (Wang et al., 1998):
  - Clinical diagnosis difficult, presentation often incomplete
  - Most consistent early diagnostic signs: Shoulder pain with ROM (flexion/abduction/external rotation), absence of pain in elbow and with forearm pronation/supination; wrist dorsiflexion pain with dorsal edema; pain with metacarpophalangeal (MCP)/proximal interphalangeal (PIP) flexion with fusiform PIP edema
  - Pain disproportionate to injury and clinical findings
  - Shoulder/hand pain preceded by rapid ROM loss
  - In this study, the most valuable clinical sign was MCP tenderness to compression with 100% predictive value, 85% sensitivity, and 100% specificity.
- **Skeletal scintigraphy (bone scan)**: Tepperman et al. (1984) and Greyson and Tepperman (1984) studied 85 consecutive poststroke hemiplegic patients with clinical features of CRPS.
  - 25% had radionuclide evidence for CRPS type I: Positive diagnosis was evident when delayed image showed increased uptake in wrist, MCP, and interphalangeal (IP) joints.
- **Stellate ganglion block**:
  - 🕮 Successful when patient develops an ipsilateral Horner syndrome (ptosis, miosis, and anhidrosis)
  - Alleviation of pain following sympathetic blockade of the stellate ganglion using local anesthetic is the gold standard for diagnosis of *sympathetically mediated* CRPS type I.

TREATMENT (Arlet & Mazieres, 1997)

- ROM exercises of the involved joint: Pain-free within 3 weeks, most <4 to 6 days with passive stretch of the involved joints.
- Corticosteroids (systemic): A large majority of patients respond to systemic steroids instituted in the acute phase of the disease. Usually prednisone in doses up to 100 to 200 mg/day or 1 mg/kg, and tapered over 2 weeks.
  - More effective in CRPS type I confirmed by bone scan than in "clinical" CRPS type I with a negative bone scan.
  - Bone scans may be useful not only in establishing the diagnosis of CRPS type I but also in identifying patients who are more likely to respond to oral steroid therapy. In a study, 90% of patients with positive bone scan findings for CRPS type I treated with corticosteroids had a good or excellent response, whereas 64% of patients without bone scan abnormalities had a poor or fair response.
  - In a recent study, 31/34 MCA stroke patients with CRPS became pain-free by 14 days after starting methylprednisolone 8 mg PO four times a day (patients treated for 2 weeks followed by 2-week taper; Braus et al., 1994).
- Medications:
  - Nonsteroidal anti-inflammatory drugs (NSAIDs)—analgesia
  - Tricyclic antidepressants (TCAs)
  - Bisphosphonates
  - Calcitonin

  - Anticonvulsants (e.g., gabapentin, carbamazepine)
  - Alpha-adrenergic blockers (e.g., clonidine, prazosin)
  - Beta-blockers (e.g., propranolol, pindolol)
  - Calcium channel blockers (e.g., nifedipine)
  - Topical capsaicin
- Modalities:
  - Edema control measures
  - Transcutaneous electrical nerve stimulation (TENS)
  - Desensitization techniques
  - Contrast baths
  - Ultrasound
- Injections:
  - Intra-articular corticosteroid injections
  - Local injections (procaine, corticosteroid)
  - Sympathetic stellate ganglion blocks may be diagnostic as well as therapeutic
- Other procedures:
  - Radiofrequency or surgical sympathectomy

Shoulder Subluxation

- Clinically presents with a palpable gap between the acromion and humeral head. Characterized by subluxation of the humeral head from the glenoid fossa inferiorly.
- Conflicting evidence as to whether subluxation is correlated with shoulder pain.
- **Etiology:** Unknown but may be due to changes in the mechanical integrity of the glenohumeral joint.
- **Pathogenesis:** Factors that are thought to be related to shoulder subluxation include:
  - Angulation of the glenoid fossa
  - Influence of the supraspinatus muscle on the humeral head sitting
  - Support of the scapula on the rib cage
  - Contraction of the deltoid and rotator cuff muscles on the abducted humerus
- A number of recent studies failed to show any correlation between shoulder subluxation and pain.
- May be a correlation between shoulder pain and decrease in arm external rotation.
- *Basmajian principle*: Decreased trapezius tone → the scapula rotates and the humeral head subluxes from the glenoid fossa.

TREATMENT

- Shoulder sling use is controversial. Routine use of sling for the subluxed shoulder (or for shoulder pain) is not indicated.
  - 🕮 Friedland (1975): Slings do not prevent/correct subluxation and are not necessary to support pain-free shoulder.
  - 🕮 Hurd et al. (1974): No appreciable difference in shoulder ROM, subluxation, or shoulder pain in patients with or without slings.
  - **Pros:** May be used when patient ambulates to support extremity (may prevent UE trauma, which in turn may cause increase in pain or predispose to development of CRPS).
  - **Cons:** May encourage contractures in shoulder adduction/internal rotation or elbow flexion (flexor synergy pattern).
- Other widely used treatments for shoulder subluxation:
  - FES
  - Arm board, arm trough, lapboard—used in poor UE recovery and primary wheelchair users
  - Arm board may overcorrect subluxation
  - Overhead slings—prevents hand edema (may use foam wedge on arm board)

- There is insufficient evidence to conclude whether slings and wheelchair attachments prevent subluxation, decrease pain, increase function, or adversely increase contracture in the shoulder after stroke. There is some evidence that strapping the shoulder delays the onset of pain but does not decrease it, nor does it increase function or adversely increase contracture (Ada et al., 2005).

PREVENTION

- Subluxation may be prevented by combining the early reactivation of shoulder musculature (specifically supraspinatus and post- and middeltoid) with the provision of FES or a passive support of the soft tissue structures of the glenohumeral joint (e.g., arm trough).

Bicipital Tendinitis

- Chronic pain in anterior shoulder, pain in abduction/external rotation, painful over bicipital groove.
- Yergason's test: With the elbow flexed at 90°, and while the forearm is pronated, the patient supinates the forearm, flexes the elbow, and externally rotates the humerus while the examiner resists these movements and pulls downward on the elbow. The test result is positive if the patient experiences pain over the bicipital groove or if the tendon pops out of the groove (see Chapter 4, Musculoskeletal Medicine).
- Greatest excursion of long head biceps from flexion/internal rotation, to elevation/abduction, depression/external rotation/extension.
- May progress to adhesive capsulitis (frozen shoulder).
- **Diagnosis:** Positive clinical exam findings and imaging studies. May be confirmed with diagnostic tendon sheath injection of lidocaine, but caution should be used with steroid injection of the tendon sheath due to risk of rupture.

Rotator Cuff Tear, Shoulder Impingement Syndrome, and Adhesive Capsulitis (Table 1–8)

- All can be potential causes of poststroke shoulder pain.
- See Chapter 4, Musculoskeletal Medicine for more details on these topics.

🕮 Brachial Plexus/Peripheral Nerve Injury

- **Etiology:** "Traction" injury to the plexus/nerve
- **Diagnosis:**
  - Atypical functional return, segmental muscle atrophy, finger extensor contracture, delayed onset of spasticity
  - Electrodiagnostic studies (EMG)—lower motor neuron findings of a brachial plexopathy
- **Treatment:**
  - Proper bed positioning to prevent patient from rolling onto paretic arm, trapping it behind the back or through bed rails and causing traction stress
  - ROM to prevent contracture while traction avoided
  - 45° shoulder abduction sling for nighttime positioning
  - Sling for ambulation to prevent traction by gravity
  - Armrest in wheelchair as needed
- **Prognosis**: May require 8 to 12 months for reinnervation

Heterotopic Ossification

- Infrequent in stroke but may be seen in elbow or shoulder
- Occurs only on extensor side of elbow
  - No problems in pronation/supination since proximal radioulnar joint not involved
- **Treatment:** Joint mobilization/ROM, etidronate disodium

Dependent Edema

- May be treated with compression glove, foam wedge, pneumatic compression, retrograde massage, and arm elevation

TABLE 1–8 Poststroke Shoulder Pain

	INFERIOR SUBLUXATION	ROTATOR CUFF TEAR	CRPS TYPE I (RSD)	ADHESIVE CAPSULITIS (FROZEN SHOULDER)	IMPINGEMENT SYNDROME	BICIPITAL TENDINITIS
Exam	• Acromiohumeral separation • Flaccid	• Positive abduction test • Positive empty can test • Positive drop arm test • Flaccid or spastic	• MCP compression test • Skin changes color • Flaccid or spastic	• External rotation <15° • Early decrease in scapular motion • Spastic	• Pain with abduction of 70°–90° • Positive Neer's test; end-range pain with forward flexion • Positive Hawkins sign • Usually spastic	• Positive Speed's/ Yergason's test • Flaccid or spastic
Diagnostic test	• X-ray in standing position • Scapular plane view	• X-ray • Arthrogram • MRI • Subacromial injection of lidocaine	• Triple-phase bone scan • Stellate ganglion block	• Arthrogram	• Subacromial injection of lidocaine	
Treatment	• Sling when upright • FES	• Steroid injection • PT/ROM • Possible surgical repair • Reduction of internal rotator cuff tone	• Oral corticosteroids • Stellate ganglion block	• PT/ROM • Debridement manipulation • Subacromial/ GH steroid injections • Intra-articular steroids • Oral steroids • Reduction of internal rotator cuff tone	• PT/ROM • Scapular mobilization • Subacromial steroid injection • Reduction of internal rotator cuff tone	

CRPS I, complex regional pain syndrome type I; FES, functional electrical stimulation; GH, glenohumeral; MCP, metacarpophalangeal; RSD, reflex sympathetic dystrophy; PT/ROM, physical therapy/range of motion.
Source: Adapted from Black-Schaffer RM, Osberg JS. Return to work after stroke: development of a predictive model. *Arch Phys Med Rehabil*. 1990;71:285–290.

OTHER STROKE REHABILITATION ISSUES

Spasticity

- For a more detailed discussion, see Spasticity section in Chapter 12, Associated Topics.
- Spasticity is defined as a velocity- and muscle length-dependent increase in resistance to externally imposed muscle stretch, resulting from hyperexcitable descending excitatory brainstem pathways and from the resultant exaggerated stretch reflex responses (Li et al., 2021).
- Usually follows classic UE flexor and LE extensor patterns.

- Spasticity usually develops days to weeks after a stroke.
- **Treatment:**
  - Noninvasive: Stretching, splints/orthosis, serial casting, electrical stimulation, cold modalities
  - Although widely done in clinical practice, the use of medications (e.g., benzodiazepines, baclofen, dantrolene, clonidine, tizanidine) in stroke patients still lacks scientific evidence to support efficacy. These drugs appear to have modest effects on the hypertonicity and posturing associated with stroke; side effects limit their usefulness (Lindsay et al., 2016).
  - Chemoneurolysis:
    - Botulinum toxin: May be particularly useful in focal control spasticity (e.g., wrist and finger flexors in the UE; ankle invertors in the LE).
    - Phenol/alcohol (neurolytic agents): May be helpful in treating spasticity of large muscle groups (e.g., hip adductors and extensors, the pectorals, lats, and biceps). Use is limited by adverse effects (e.g., pain during injection, postinjection dysesthesia/chronic pain, and vascular injection).
  - Intrathecal baclofen (ITB) pump (Park & Song, 2024):
    - The use of ITB pumps for spasticity management after stroke was found to improve spasticity and gait speed.
    - Potentially can treat severe spasticity unresponsive to conventional spasticity treatment.
    - Must consider individual patient characteristics and conditions when contemplating ITB injection for spasticity intervention.
  - Surgical procedures:
    - Uncommonly used in stroke, probably because of the expected decrease in survival and increase in rate of medical comorbidities.
    - May be useful in selected cases when specific goals are pursued (e.g., increase in function, improvement in hygiene, decrease in pain).

Deep Vein Thrombosis

- Common medical complication after stroke, occurring in 20% to 75% of untreated stroke survivors.
- 60% to 75% in affected extremity, 25% are proximal DVT; 1% to 2% pulmonary embolism (PE).
- **Diagnosis:** Usually can be made using noninvasive techniques.
  - Ultrasonography
  - Impedance plethysmography
  - Contrast venography reserved for cases with inconclusive results
  - D-dimer assays (a cross-linked fibrin degradation product): May be useful as a screening tool for DVT in stroke patients
- **Recommended prophylaxis regimens** (Winstein et al., 2016):
  - In ischemic stroke, prophylactic-dose subcutaneous heparin (low-molecular-weight over unfractionated) should be used for the duration of the acute and rehabilitation hospital stay or until the stroke survivor regains mobility.
  - In ICH, it may be reasonable to use prophylactic-dose subcutaneous heparin (low-molecular-weight over unfractionated) started between days 2 and 4 over no DVT prophylaxis.
  - In ischemic stroke and ICH, it may be reasonable to use intermittent pneumatic compression over no prophylaxis during the acute hospitalization.
  - In ischemic stroke and ICH, it is not useful to use elastic compression stockings.

Bladder Dysfunction (also see Chapter 7, Spinal Cord Injuries, Bladder Dysfunction section)

- Incidence of urinary incontinence is 50% to 70% during the first month after stroke and 15% after 6 months (similar to general population incidence of approximately 17%).

- Incontinence may be caused by CNS damage itself (i.e., neurogenic bladder), urinary tract infection (UTI), impaired ability to transfer to toilet, or impaired mobility, confusion, communication disorder/aphasia, and cognitive-perceptual deficits that result in lack of awareness of bladder fullness.
- Types of voiding disorders: Areflexia, uninhibited spastic bladder (with complete/incomplete emptying), and outlet obstruction.
- **Treatments**:
  - Treatment of possible underlying medical causes (e.g., UTI)
  - Regulation of fluid intake
  - Transfer and dressing-skill training
  - Patient and family education
  - Medications (if no improvement with conservative measures)
- Antimuscarinic agents, such as oxybutynin, tolterodine, fesoterodine, darifenacin, solifenacin, trospium, and imipramine, prevent acetylcholine release from parasympathetic nerves by binding to receptors on the detrusor muscle, thus assisting in bladder storage. Side effects include dry mouth, blurry vision, constipation, and increased risk of exacerbating altered cognition (particularly with use of oxybutynin or tolterodine; Duong et al., 2021).
- Bethanechol, a cholinergic agonist, increases detrusor activity, but there is lack of controlled clinical trials to support frequent use of this medication. Cholinergic side effects include flushing, sweating, diarrhea, hypersalivation, and abdominal cramps (Agarwal, 2021).
- Adrenergic antagonists, including phenoxybenzamine, prazosin, terazosin, doxazosin, tamsulosin, and silodosin, inhibit smooth muscle activity in the prostate and at the bladder neck. Common side effects are postural hypotension, abnormal ejaculation, nasal congestion, fatigue, and headache. Aforementioned side effects are more common with terazosin and doxazosin (Agarwal, 2021). Recent trials suggest that tamsulosin improves bladder storage and decreases the symptoms of autonomic dysreflexia in spinal cord injury patients.
- For patients with mild stress incontinence, adrenergic agonists, like ephedrine and mirabegron, increase urethral resistance. However, they can increase BP and heart rate and cause dizziness (Yuanzhuo et al., 2022).
- Estrogen administration can help restore or maintain urethral submucosa tissue in women who have stress incontinence.
- Baclofen, tizanidine, diazepam, and dantrolene sodium are examples of muscle relaxants for skeletal muscle spasticity.
- For detrusor overactivity refractory to medications, botulinum toxin type A injections and a bladder stimulator are FDA-approved therapies that may be considered.
  - Timed bladder-emptying program
  - Remove indwelling catheter and perform postvoid residuals (PVRs)
  - Intermittent catheterization (IC)
  - Urodynamics evaluation

Bowel Dysfunction

- Incidence of bowel incontinence in 31% stroke patients
- Patient unable to inhibit urge to defecate → incontinence
- Incontinence usually resolves within the first 2 weeks. Persistence may reflect severe brain damage.
- Decrease in bowel continence may be associated with infection, inability to transfer to toilet or to manage clothing, and communication impairment/inability to express toileting needs.
- **Treatment:** Treat underlying causes (e.g., bowel infection, diarrhea); timed toileting schedule, training in toilet transfer, and communication skills

Impairment of Intestinal Peristalsis-Constipation

- **Management:** Adequate fluid intake/hydration, diet modification (e.g., increase dietary fiber), bowel management (stool softeners, stool stimulants, and suppositories), and allow commode/bathroom privileges

Dysphagia

- Dysphagia (difficulty swallowing) has an overall prevalence of 25% to 65% in stroke patients and occurs in the following regions (Gordon et al., 1987):
  - 67% of brainstem strokes
  - 28% of all left hemispheric strokes
  - 21% of all right hemispheric strokes
- More common in bilateral hemisphere lesions than in unilateral hemisphere lesions
- More common in large-vessel than in small-vessel strokes
- Delayed pharyngeal swallow is the most common cause (Veis & Logemann, 1985).

🕮 DIAGNOSIS OF DYSPHAGIA

- **Bedside swallowing evaluation**
  - A minimally invasive evaluation procedure that helps determine whether dysphagia is present
  - Includes a medical history review for risk
  - May evaluate gag reflex or pharyngeal sensation
  - Observes for overt signs of cough or other difficulty during swallowing trials
  - Determines whether it is safe to feed a patient orally for purposes of nutrition, hydration, and administering medications
  - Determines whether a patient requires further swallowing assessments
  - Determines whether a patient requires referral for nutritional or hydrational support
- **Videofluorographic swallowing study (VFSS)**
  - Also called videofluoroscopic swallowing evaluation and modified barium swallow (MBS)
  - 🕮 "Gold standard" for evaluation and treatment of dysphagia
  - Different amounts and consistencies of solids and liquids mixed with barium are swallowed while visualizing the anatomy of swallowing.
  - If abnormal swallowing is identified, the clinician determines the physiologic causes for swallowing breakdowns.
  - Compensatory techniques, postures, maneuvers, sensory enhancements, and bolus modifications are attempted to determine methods of safe feeding.
  - If no method of safe feeding can be determined, alternative methods of feeding must be considered.
- 🕮 **Fiberoptic endoscopic evaluation of swallowing (FEES)**
  - Used as a comprehensive functional evaluation of the pharyngeal stage of swallowing
  - Visualizes anatomic structures that might cause potential bolus obstruction and natural bolus flow and containment
  - Different amounts and consistencies of solids and liquids are swallowed while visualizing the anatomy of swallowing.
  - Evaluates swallowing physiology, coordination, and associated events (e.g., obstruction, absence of natural protective anatomy, depth and duration of bolus passage into the pharynx/larynx)
  - Evaluates the reaction to the presence of residue, penetration, and/or aspiration (e.g., reduction in percent residue, effectiveness of cough, and expectoration of material from the airway)

🕮 Aspiration

- Aspiration is the entry of a substance through the vocal folds (true vocal cords) into the trachea.
- 🕮 Aspiration is missed on bedside swallowing evaluations in 40% to 60% of patients (i.e., silent aspiration).
- It can be reliably diagnosed on a VFSS. Aspiration will show penetration of contrast material below the true vocal cords.
  - Aspiration has been found to occur in 40% to 70% of stroke patients on VFSS.

- Predictors of aspiration on VFSS include:
  - Delayed initiation of the swallow reflex
  - Decreased pharyngeal peristalsis
- 🕮 Predictors of aspiration on bedside swallowing exam:
  - Abnormal cough
  - Cough after swallow
  - Dysphonia
  - Dysarthria
  - Abnormal gag reflex
  - Voice change after swallow (wet voice; Aronson, 1990)

🕮 Aspiration Pneumonia

- Risk factors for development of pneumonia secondary to aspiration include:
  - Decreased level of consciousness
  - Tracheostomy
  - Emesis
  - Reflux
  - Nasogastric tube (NGT) feeding
  - Dysphagia
  - Prolonged pharyngeal transit time
- As dysphagia is a frequent and potentially serious complication of stroke (because of the risk of aspiration), careful bedside swallowing evaluation should be performed in all patients before oral feeding is started.
- If a patient is believed to be at high risk of recurrent aspiration after bedside and/or videofluorographic evaluation, the patient should be kept NPO and enterally fed, initially via NGT and then via G- or J-tube if long-term enteral feeding is required.

🕮 THE FOUR PHASES OF SWALLOWING (TABLES 1–9 TO 1–12)

1. Oral preparatory phase
2. Oral phase
3. Pharyngeal phase
4. Esophageal phase

TABLE 1–9 Oral Preparatory Phase

Voluntary vs. reflex	• Voluntary
Phase duration	• Variable: Voluntary phase with duration influenced by consistency of material ingested, number of times person chews, etc.
Hallmarks of this phase	• Preparation of bolus
Phase requires	• Tension in the labial and buccal musculature to close the anterior and lateral sulci • Rotary jaw (circular) motion for mastication/grinding • Lateral tongue movement to position food on the teeth during mastication (tongue moves food back to teeth) • Depression and forward movement of the soft palate to seal the oral cavity posteriorly and widen the nasal airway • Saliva
Problems seen in this phase	• Drooling • Pocketing

TABLE 1–10 Oral Phase

Voluntary vs. reflex	• Voluntary
Phase duration	• Lasts usually <1 second
Hallmarks of this phase	• Tongue that elevates and occludes the anterior oral cavity and compresses the bolus toward the oropharynx • Contraction of the palatopharyngeal folds • Elevation of the soft palate
🕮 **Phase requires**	• Tension in the labial and buccal musculature to close the anterior and lateral sulci • Anterior–posterior tongue movement to transport the bolus to the pharynx • Soft palate elevation and velopharyngeal port closure (also seen in the pharyngeal phase)—to close off the nasal cavity and prevent regurgitation into the nasopharynx
Problems seen in this phase	• Drooling • Pocketing • Head tilt

TABLE 1–11 Pharyngeal Phase

Voluntary vs. reflex	• Reflex
Phase duration	• Lasts approximately 0.6–1 second
Hallmarks of this phase	• Bolus propelled from mouth to esophagus • Aspiration is *most* likely to occur during this phase • With initiation of pharyngeal phase, inhibition of breathing occurs to prevent aspiration
🕮 **Phase requires**	• Soft palate elevation and velopharyngeal port closure (also seen in the oral phase)—to close off the nasal cavity and prevent regurgitation into the nasopharynx • Laryngeal elevation, with forward movement of the hyoid bone and folding of the epiglottis to protect the airway • Adduction of the ventricular and true vocal folds to protect the airway • Coordinated *pharyngeal constriction* and *cricopharyngeal* (upper esophageal sphincter) *relaxation*—to facilitate bolus transport into the esophagus
Problems seen in this phase	• Food sticking, choking and coughing, aspiration, wet/gurgling voice, nasal regurgitation

TABLE 1–12 Esophageal Phase

Voluntary vs. reflex	• Reflex
🕮 **Phase duration**	• Longest phase—lasts 6–10 seconds
Hallmarks of this phase	• Bolus pass from pharynx → esophagus → stomach • Esophageal clearance is assisted by gravity but requires relaxation of the gastroesophageal sphincter
Phase requires	• Cricopharyngeal muscle contraction • Coordinated peristalsis and LES relaxation
Problems seen in this phase	• Heartburn, food sticking

LES, lower esophageal sphincter.

Treatment of Dysphagia/Prevention of Aspiration

- **Oral feeding**:
  - Modifications of diet consistency (thickened fluids, pureed or soft foods in smaller boluses) if the patient is able to tolerate PO intake without evidence of aspiration.
- **Nonoral feeding (NPO):**
  - Clear contraindication for oral feeding is pulmonary pathology due to aspiration in the presence of documented airway contamination.
  - NPO indicated in patients at high risk of aspiration because of reduced alertness, reduced responsiveness to stimulation, absent swallow, absent protective cough, and difficulty handling secretions, or when there is significant reduction of oral pharyngeal and laryngeal movements.
  - NPO is disadvantageous in treating dysphagia because swallowing itself is the best treatment.
- Changes in posture and head position
- Elevation of the head of the bed
- Feeding in the upright position
- **Compensatory strategies**:
  - 🕮 **Chin tuck**: Provides airway protection by preventing entry of liquid into the larynx, probably by facilitating forward motion of the larynx. Chin tuck also decreases the space between the base of the tongue and the posterior pharyngeal wall, and so creates increased pharyngeal pressure to move the bolus through the pharyngeal region.
  - 🕮 **Head rotation**: Closes ipsilateral pharynx, forces bolus into contralateral pharynx, and decreases cricopharyngeal pressures. Turn the head to the paretic side.
  - **Head tilt**: Uses gravity to guide bolus into ipsilateral pharynx.
  - **Supraglottic swallow**: Concomitant breath holding and swallowing closes the vocal folds to protect the trachea.
  - **Super supraglottic swallow**: Adds Valsalva maneuver to maximize vocal fold closing.
  - **Mendelsohn maneuver**: Patient voluntarily holds the larynx at its maximal height to lengthen the duration of the cricopharyngeal opening.
- Other treatment modalities (inconclusive evidence of long-term efficacy in dysphagia):
  - Thermal stimulation (to sensitize the swallowing reflex)
  - Oral/motor exercises (to improve tongue and lip strength, ROM, velocity, and precision, and vocal fold adduction)

Complications of Dysphagias

- Dehydration
- Malnutrition was found in 49% of patients admitted to rehabilitation in a recent study and associated with a prolonged length of stay and slower rate of functional gains (Finestone et al., 1995).
- Malnourished patients also found to have higher stress reaction and higher frequency of infection and decubitus ulcers.

Recovery of Dysphagia in Stroke

- Few studies available on recovery of dysphagia in stroke.
- 🕮 Ickenstein et al. (2012): Determined Swallowing Functional Communication Measure (FCM) and Penetration-Aspiration Scale (PAS) scores in subjects at 72 hours poststroke and tracked outcomes at 90 days poststroke
  - At 72 hours, subjects with FCM scores of 1 to 3 and PAS scores of 5 to 8 were 11.8 times less likely to be orally fed at 90 days poststroke.
  - 86% of subjects at 72 hours poststroke with FCM scores of 4 to 6 and PAS scores of 2 to 4 (no confirmed signs of aspiration) developed only mild swallowing deficits by 90 days poststroke.

- Wilkinson et al. (2002): 12-month retrospective cohort study of 186 teaching hospital inpatients
  - Subjects 7 days poststroke who tolerated grade 1 fluids or thinner, or who tolerated a modified soft diet or better: None needed a percutaneous endoscopic gastrostomy (PEG) device.
  - Subjects needing PEG 14 days poststroke: 50% could not tolerate grade 3 thickened fluids; 52% could not tolerate a pureed diet.
  - No subject intolerant of grade 2 thickened fluids 7 or 14 days poststroke could tolerate a normal diet and fluids by day 28.
  - Of subjects tolerating grade 3 thickened fluids at day 7, 36% tolerated a normal diet at day 28.
  - PEG placement should be considered in people unable to tolerate grade 3 thickened fluids or pureed diet 14 days poststroke. However, even in these groups, half recovered sufficiently to manage oral feeding.
- Gresham (1990) reports findings regarding 53 patients in a swallowing program poststroke.
  - 85% were on full oral nutrition at discharge.
  - 17% could not drink thin liquids safely.
  - 8% could not adequately maintain cohesive bolus of varied texture.
- Logemann (1991): Recovery of swallowing function in most brainstem strokes occurs in the first 3 weeks poststroke.
- Within 7 days, swallowing dysfunction recovers for about half of patients. At 6 months, only 11% to 13% have persistent swallowing dysfunction (Stein et al., 2015).
- 🕮 **Nasal speech**: Hypernasality caused by partial or complete failure of soft palate to close off the nasal cavity from the oral cavity or by incomplete closure of the hard palate. Uplifting the soft palate prevents nasal speech (speech abnormally resonated in the nasal cavities).

APHASIA (TABLE 1–13)

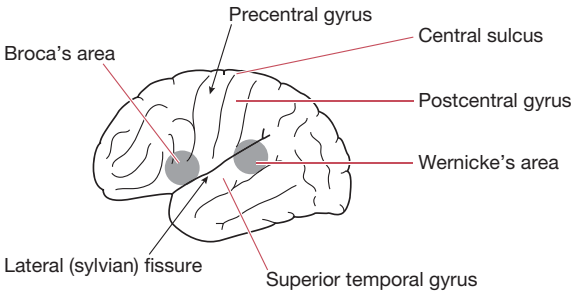
- Aphasia is an impairment of the ability to utilize language due to an injury to the brain. It is characterized by paraphasias, word-finding difficulties, and impaired comprehension. Other common but not obligatory features are disturbances in reading and writing, nonverbal constructional and problem-solving difficulty, and impairment of gesturing.

TABLE 1–13 Types of Aphasias

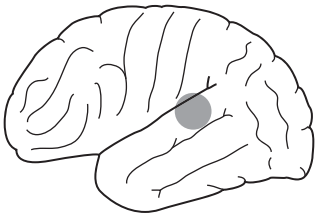
FLUENT APHASIA				NONFLUENT APHASIA			
+ COMPREHENSION		– COMPREHENSION		+ COMPREHENSION		– COMPREHENSION	
REPETITION ⇓		*REPETITION* ⇓		*REPETITION* ⇓		*REPETITION* ⇓	
+	–	+	–	+	–	+	–
NAMING	Conduction	Transcortical sensory	Wernicke's	Transcortical motor	Broca's	Mixed transcortical	Global
+	–						
Normal	Anomia						

FLUENT APHASIAS	NONFLUENT APHASIAS
Wernicke's Transcortical sensory Conduction Anomia	Broca's Transcortical motor Global Mixed transcortical

🕮 ANATOMIC LOCATION OF MAJOR SPEECH AREAS



WERNICKE'S APHASIA

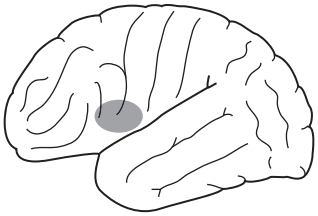


🕮 Location: Posterior part of superior (first) temporal gyrus of the dominant (usually left) hemisphere

Characteristics:

- Fluent speech (normal rate/speed)
- Impaired comprehension
- Word deafness, difficulty in reading (alexia) and writing (agraphia)
- Marked paraphasias and neologisms

BROCA'S APHASIA (REMEMBER "BROKEN" SPEECH)

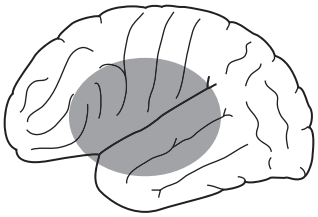


🕮 Location: Posterior–inferior frontal lobe (third frontal convolution) of dominant (usually left) hemisphere → anterior to motor cortex areas that supply the tongue, lips, and larynx

Characteristics:

- Nonfluent speech (telegraphic)
- Impaired repetition
- Preserved comprehension
- Paraphasias and articulatory errors or struggle

GLOBAL APHASIA

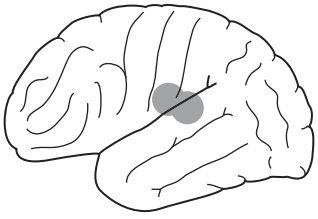


🕮 Location: Vary in size and location but usually involve distribution of the left middle cerebral artery (entire perisylvian region)

Characteristics:

- Ranges from mutism (nonfluent) to total repetitive jargon or neologistic output (fluent but incomprehensible speech)
- Poor comprehension and repetition

ANOMIC APHASIA

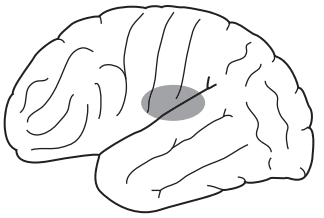


🕮 Location: Temporoparietal injury, angular gyrus

Characteristics:

- Fluent, essentially good comprehension and repetition
- Decreased output of nouns
- Word-finding difficulties
- Alexia and agraphia may be present

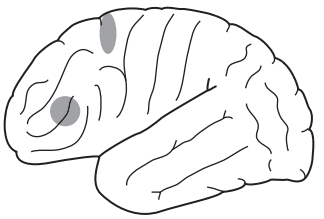
CONDUCTION APHASIA



📖 Location: Lesion of the parietal operculum (arcuate fasciculus) or insula or deep to the supramarginal gyrus (usually left side)
Characteristics:
- Normal rate of speech (fluent)
- Preserved comprehension
- Impaired repetition
- Literal paraphasias with "targeting" of words (until getting the right one)

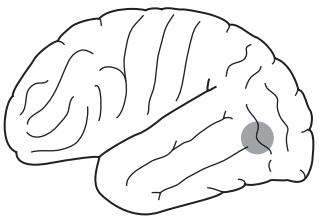
Note: **Arcuate fasciculus** is a band of white matter running deep to the supramarginal gyrus and insula that joins Broca's and Wernicke's areas.

TRANSCORTICAL MOTOR APHASIA



Location: Frontal lobe, anterior or superior to Broca's area or in the subcortical region deep to Broca's area
Characteristics:
- Reduced rate of speech, limited language output (with some fluent utterances)
- Reduced initiation and organization of speech
- Comprehension usually intact
- Preserved repetition

TRANSCORTICAL SENSORY APHASIA



Location: Watershed lesion isolating perisylvian speech structures (Broca's and Wernicke's areas) from the posterior brain; angular gyrus or posterior–inferior temporal lobe
Characteristics:
- Poor comprehension
- Fluent speech (neologisms)
- Preserved repetition (possibly echolalia)

Transcortical Mixed Aphasia

- Also known as isolation aphasia
- Lesions in border zone of frontal, parietal, and temporal areas
- Characteristics:
  - Poor comprehension
  - Nonfluent (decreased rate and initiation of speech)
  - Preserved repetition (echolalia)

Note: Language areas are anatomically clustered around the sylvian fissure of the dominant hemisphere-left hemisphere in 95% of people.

📖 Other Errors in Speech

- **Paraphasia**: Incorrect substitution of words or parts of words.
  - Literal or phonemic paraphasias: Similar sounds (e.g., "sound" for "found")
  - Verbal or semantic paraphasias: Word substituted for another from same semantic class (e.g., "fork" for "spoon")
- **Agrammatism**: Aphasia in which there is absence of grammatical structure in a sentence.
- **Anomia (anomic or nominal aphasia):** Difficulty recalling words; word-finding difficulty. Unimpaired comprehension and repetition.

- **Echolalia**: Repetition ("echoing") of words or vocalizations made by another person.
- **Circumlocution**: Roundabout way of describing a word that cannot be recalled. Often seen in conjunction with anomia.
- **Neologism**: A "new word" that is well-articulated but has meaning only to the speaker.
- **Jargon**: Well-articulated but mostly incomprehensible, unintelligible speech. Associated with Wernicke aphasia.
- **Stereotype**: Repetition of nonsensical syllables (e.g., "no, no, no") during attempts at communication.

Treatment Approaches to Aphasias

- Loss versus interference (most widely accepted approach): Views aphasia as loss of specific linguistic information (compensatory) with concept that brain damage interferes with linguistic operation (facilitatory)
- Direct versus indirect: Views aphasia as deficit of linguistic thought processes, which can be used to differentiate direct language-centered therapy from indirect content-centered therapy
- Behavioral versus psycholinguistic: Views emphasis on content, emphasis on structure of therapy
- Programmed operant: Measures obtained before and after behavior-modification procedure applied
- Programmed instruction: Involves many individual steps to reach a desired language behavior
- Augmentative and alternative communication (AAC): Use of ways to augment limited communication or alternate ways of communication (Table 1–14)

TABLE 1–14 Forms of Augmentative and Alternative Communication

UNAIDED (USING ONLY YOUR BODY WITHOUT AN EXTERNAL DEVICE)	AIDED (USING A TOOL OR DEVICE)	
No Tech	Low/Light Tech	High Tech
• Gestures • Manual signs • Facial expressions • Vocalizations • Verbalizations • Body language	• Pictures • Objects • Photographs • Writing • Communication boards/books	• SGD • Single-message devices and recordable/digitized devices • AAC software that enables dynamic symbol/language representation and that is used with some form of technology hardware (e.g., computer, tablet, smartphone)

AAC, augmentative and alternative communication; SGD, speech-generating devices.
Source: Adapted from Augmentative and alternative communication: key issues. American Speech-Language-Hearing Association. https://www.asha.org/practice-portal/professional-issues/augmentative-and-alternative-communication/.

- Specific interventions:
  - AmerInd: Sign language that uses gestures that represent objects, actions, directions, and descriptions.
  - 🕮 Melodic intonation therapy (MIT): Recruits the right hemisphere for communication by incorporating melodies or rhythms with simple statements. MIT may be useful in patients with nonfluent (Broca's) aphasia.

🕮 Poststroke Aphasia Recovery

- The greatest amount of improvement in patients with aphasia occurs in the first 2 to 3 months after the onset.
- After 6 months, there is a significant drop in the rate of recovery.
- In the majority of cases of patients with aphasia, spontaneous recovery does not seem to occur after 1 year.
- However, there are reports of improvements many years after stroke in patients undergoing therapy.

MANAGEMENT OF MEDICAL ISSUES

Poststroke Depression

- **Etiology**:
  - Organic: May be related to catecholamine depletion through lesion-induced damage to the frontal noradrenergic, dopaminergic, and serotonergic projections (Heilman et al., 2012).
  - Reactive: Grief/psychological responses for physical and personal losses associated with stroke, loss of control that often accompany severe disability, and so on.
- Prevalence of depression in stroke patients reported at approximately 40% (25%–79%); occur in similar proportion in their caregivers (Flick, 1999).
- Most prevalent 6 months to 2 years.
- A psychiatric evaluation for *Diagnostic and Statistical Manual of Mental Disorders,* Fifth Edition *(DSM-5)* criteria and vegetative signs may be a clinically useful diagnostic tool in stroke patients.
- There may be a higher risk for major depression in left frontal lesions (relationship still controversial).
- **Risk factors:** Prior psychiatric history, significant impairment in ADLs, high severity of deficits, female gender, nonfluent aphasia, cognitive impairment, and lack of social supports.
- Persistent depression correlates with delayed recovery and poorer outcome.
- **Treatment:** Active treatment should be considered for all patients with significant clinical depression.
  - Psychosocial interventional program: Psychotherapy
  - SSRIs preferred because of fewer side effects compared with TCAs; methylphenidate has also been shown to be effective in poststroke depression.
  - SSRIs and TCAs also have been shown to be effective in poststroke emotional lability.

Sexual Dysfunction

- Well-documented that the majority of older adults continue to enjoy active and satisfying sexual relationships.
- No significant changes in sexual interest or desire, but marked decline in behavior in both sexes after a stroke.
- There is a marked decline in sexual activity poststroke.
- Korpelainen et al. (1999): 192 stroke patients and 94 spouses
  - 79% of patients and 84% of spouses reported active prestroke sexual life, including intercourse at least once a month.
  - After the stroke, however, the numbers decreased, with 45% of patients and 48% of spouses reporting an active sexual life.
  - 33% of patients and 27% of spouses reported having ceased sexual intercourse.
- Decreased coital frequency associated with inability to discuss sexuality with spouse (odds ratio [OR] 18.5); general attitude toward sexuality (unimportant: OR 7.7; fairly important: OR 9.2); unwillingness to participate in sexual activity (OR 5.4).
- Other factors related to decrease in sexual activity poststroke:
  - **Emotional factors:** Fear, anxiety, and guilt; low self-esteem; and fear of rejection by partner
  - Medications that worsen sexual function: TCAs, SSRIs, antipsychotics, anticholinergics, histamine (1 and 2) blockers, gamma-aminobutyric acid (GABA) agonists (e.g., pregabalin), opioid narcotics, saw palmetto
  - **Treatment:** Supportive psychotherapy, counseling, medical consultation (e.g., urology, urogynecology) for organic causes

Seizures

- Can be classified as occurring:
  - At stroke onset
  - Early after stroke (1–2 weeks)
  - Late after stroke (>2 weeks)

- In a prospective study after first-time stroke, 2.5% of patients had seizures within 48 hours postictus.
- Seizures were associated with older age, confusion, and large parietal or temporal hemorrhages.
- Majority of seizures were generalized, tonic-clonic.
- In-hospital mortality rates are higher in patients with seizures.
- Early seizures tend not to recur. These are associated with acute metabolic derangement associated with ischemia or hemorrhage.
- Stroke patients requiring inpatient rehabilitation have a higher probability of developing seizures than the general stroke population.
- Seizures occurring >2 weeks after stroke have higher probability of recurrence.
- In a study with 77 ischemic stroke patients followed for 2 to 4 years:
  - 6% to 9% developed seizures.
  - 26% patients with cortical lesions developed seizures.
  - 2% patients with subcortical lesions developed seizures.
- **Risk factors:** Cortical lesions and persistent paresis (50%)
- **Treatment:** Choice of anticonvulsant drugs for patients with cerebral injury discussed in Chapter 2, Traumatic Brain Injury Medicine.

FACTORS THAT PREDICT MORTALITY AND FUNCTIONAL RECOVERY IN STROKE PATIENTS

Mortality Risk Factors

- In persons aged 45 to 64: 8% to 12% of ischemic strokes and 37% to 38% of hemorrhagic strokes die within 30 days (ARIC study, National Heart, Lung, and Blood Institute).
- In persons aged 65 and older: 12.6% of all strokes, 8.1% of ischemic strokes, and 44.6% of hemorrhagic strokes die within 1 month (Medicare Part B random sample).
- Mortality rates were higher after hemorrhagic stroke (67.9%) than after ischemic stroke (57.4%; Benjamin et al., 2018).
- Mortality in the first year after stroke: 25% to 40%
- Risk of another stroke within the first year: 12% to 25%

RISK FACTORS FOR ACUTE STROKE MORTALITY—30-DAY MORTALITY

- Stroke severity
- Decreased level of consciousness
- Diabetes mellitus
- Cardiac disease
- EKG abnormalities
- Old age
- Delay in medical care
- Elevated blood sugar in patients without diabetes
- Brainstem involvement
- Hemorrhagic stroke
- Admission from nursing home

Risk Factors for Disability After Stroke (Kelley-Hayes et al., 2003)

- As stroke mortality has declined in the last few decades, the number of stroke survivors with impairments and disabilities has increased.
- There are 300,000 to 400,000 stroke survivors annually.
- 50% had some hemiparesis.
- 30% were unable to walk without some assistance.
- 26% were dependent in ADLs.
- 19% had aphasia.
- 35% had depressive symptoms.
- 26% were institutionalized in a nursing home.

RISK FACTORS FOR DISABILITY AFTER STROKE

- Severe stroke (minimal motor recovery at 4 weeks)
- Decreased level of consciousness
- Diabetes mellitus
- Cardiac disease
- EKG abnormalities
- Old age
- Delay in medical care
- Delay in rehabilitation
- Bilateral lesions
- Previous stroke
- Previous functional disability
- Poor sitting balance
- Global aphasia
- Severe neglect
- Sensory and visual deficits
- Impaired cognition
- Incontinence persisting >1–2 weeks

🕮 NEGATIVE RISK FACTORS FOR RETURN TO WORK POSTSTROKE (Wang et al., 2014)

- Most consistent negative predictor: Stroke severity (i.e., as assessed by the degree of residual disability such as weakness and neurologic deficit or impairments [speech, cognition, apraxia, agnosia])
- Other factors to be taken into consideration: Number of working years remaining until retirement, depression, medical history, occupation
- Stroke survivors who are flexible and realistic in their vocational goals and emotionally accept their disabilities appear more likely to return to work.
- In addition, the following criteria should also be considered regarding negative risk factors for return-to-work poststroke (Black-Schaffer & Osberg, 1990):
  - Low score on Barthel index at the time of rehabilitation discharge
    - Barthel index: Functional assessment tool that measures independence in ADLs on 0 to 100 scale (www.mdcalc.com/calc/3912/barthel-index-activities-daily-living-adl)
    - The Barthel Index has ten activities that are scored individually from 0-10 or 0-15, totaling 0-100, with 100 being most independent, and 0 being most dependent. The 10 activities they assess are bathing, feeding, grooming, dressing, bladder, bowels, toilet use, transfers (from bed to chair and back), mobility on even surfaces, and stairs.
  - Prolonged rehabilitation length of stay
  - Aphasia
  - Prior alcohol abuse

THE BARTHEL ADL INDEX: GUIDELINES

1. The index should be used as a record of what a patient does, not as a record of what a patient could do.
2. The main aim is to establish degree of independence from any help, physical or verbal, however minor and for whatever reason.
3. The need for supervision renders the patient not independent.
4. A patient's performance should be established using the best available evidence. Asking the patient, friends/relatives, and nurses are the usual sources, but direct observation and common sense are also important. However, direct testing is not needed.
5. Usually the patient's performance over the preceding 24 to 48 hours is important, but occasionally longer periods will be relevant.
6. Middle categories imply that the patient supplies over 50% of the effort.
7. Use of aids to be independent is allowed.

Source: From Mahoney FI, Barthel D. Functional evaluation: the Barthel Index. *Md State Med J*. 1965;14:56–61, with permission.

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TRAUMATIC BRAIN INJURY

2

Elie Elovic, MD • Edgardo Baerga, MD • Sara J. Cuccurullo, MD • Christine Greiss, DO • Matthew Moore, DO • Yi (Dan) Zhou, MD

INTRODUCTION

EPIDEMIOLOGY

- Trauma is the leading cause of death in people ages 1 to 44, and more than one-half of these deaths are due to brain trauma. Traumatic brain injury (TBI) is arguably the primary cause of neurologic mortality and morbidity in the United States.
  - In 2013, there were approximately 2.5 million TBI-related ED visits, 282,000 hospitalizations, and 56,000 deaths (Taylor et al., 2017).
  - In 2021, there were approximately 214,000 TBI-related hospitalizations and 69,000 deaths (Centers for Disease Control and Prevention [CDC], 2021).
  - Age distribution is bimodal.
  - Peak ages: 0 to 5 years, with second peak in the elderly (age 65 and older); older group has a higher mortality rate.
- Male-to-female ratio → 2.5:1.
- Violence/assault is the second most common cause of TBI in young adults. People ages 15 to 34 years have TBI-related hospitalizations, ED visits, and death most commonly from motor vehicle crashes (Taylor et al., 2017).
- Populations at risk for TBI include racial and ethnic minorities, military personnel, people who experience homelessness, survivors of intimate partner violence, those in detention facilities, and those in rural areas (CDC, 2024).
- Ethyl alcohol (ETOH) use is clearly related to TBI.
  - Studies estimate that 30% to 50% of patients were intoxicated at the time of TBI, with greater estimates for patients injured in motor vehicle accidents (MVAs) and assaults (Weil et al., 2018).

Centers for Disease Control and Prevention Surveillance Report of 2018 to 2019 Data

- Ages 0 to 17 years: The most common causes of TBI-related deaths were motor vehicle crashes and homicide.
- Ages 15 to 34 years: TBI-related deaths due to unintentional motor vehicle crashes were highest in this age group.
- Age ≥75 years: Unintentional falls leading to TBI-related deaths are highest in this age group.

Traumatic Brain Injury Model System National Database Statistics (1989–2022)

- The TBI Model System database includes data from specific centers on the outcomes and recovery of persons with TBI who had coordinated care in acute neurotrauma and inpatient rehabilitation settings up to 30 years after injury. It currently includes 16 TBI Model System centers (Model Systems Knowledge Translation Center, 2024).
- Sex: Males > females and account for 74% of TBIs.
- Age distribution:
  - 16 to 25: 30%
  - 26 to 35: 18%
  - 36 to 45: 17%
  - 46 to 55: 14%
  - 56 to 65: 9%
  - >66: 12%
- Race: Caucasian (66%) > African American (18%) > Hispanic (11%) > Asian/Pacific Islander (3%) > Native American (1%)
- Marital status:
  - Not married (68%)
  - 🕮 Single (46%) > married (33%) > divorced (16%) > widowed/separated (5%)
- Education: 64% high school level or less
- Employment: 61% employed at time of injury
- 🕮 Alcohol-related injuries: 46%. Since this data item is often missing, this might understate the problem, as other data report >50% rate and increased risk of recurrence.

National Center for Injury Prevention and Control Statistics (CDC, 2015; Taylor et al., 2017)

- **Prevalence:** There are currently between 3.2 and 5.3 million people in the United States living with TBI-related disabilities (CDC, 2015).
  - Incidence: In 2013, there were 2.8 million TBI-related ED visits, hospitalizations, and deaths (Taylor et al., 2017):
    - Approximately 56,000 deaths related to TBI
    - Approximately 282,000 hospitalizations related to TBI
    - 2.5 million ED visits that are not admitted related to TBI
- *Deaths*: Improved acute medical care and injury prevention strategies have led to a steady decline in the incidence of TBI mortalities, with a 30-day mortality of 30%.
- *Hospitalizations*: Hospitalization rates have been steady over the last 30 years.
  - Again, highest in those over 65 years old.
- *ED visits*: Nonadmissions outnumber TBI admissions by four times.
  - The rate of ED visits for TBI is greatest in those between the ages 0 and 4 years.
- **Severity:** 90% of injuries are classified as mild. Even among those hospitalized, 75% will have a Glasgow Coma Scale (GCS) >13.
- **Cost:** Total economic impact of TBI in the United States in 2010 was approximately $76.5 billion: $12 billion in lifetime medical costs and $55 billion in productivity losses.

Mortality in Traumatic Brain Injury

- Mortality rate in TBI: 45.2 per 100,000 per year
  - Rates of TBI-related deaths per 100,000 population were highest among adults aged ≥75 years (2018–2019).
- Mortality in males is three to four times higher than in females (2018–2019).
- 🕮 The single most common cause of death and injury in automobile accidents is ejection of the occupant from the vehicle (Spitz & Fisher, 1991).
- Suicide and unintentional falls were the most common mechanisms of injury contributing to a TBI-related death. Suicide accounted for 34.7% and unintentional falls 28% of all TBI-related deaths in 2017 (CDC Traumatic Brain Injury and Concussion, accessed August 2023).

- There has been a change in trends from the 1990s to 2000s in TBI mortality:
  - Decrease in deaths secondary to MVA but increase in injuries (and deaths) due to firearms/violence
- Study of TBI deaths from 1979 to 1992 (Sosin et al., 1996):
  - Average of 52,000 deaths per year in the United States secondary to TBI (Faul et al., 2010)
  - Decline in overall TBI-related deaths of 22% from 1979 to 1992. Reasons are unknown but may be related to vehicles being equipped with airbags, increased use of seatbelts, vehicle safety improvement features, roadway safety improvements, and so on.
  - 25% decline in MVA-related deaths
  - 13% increase in firearm-related deaths
  - Intentional self-harm was the leading cause of death for persons 25 to 64 years of age.
  - Motor vehicle crashes were the leading cause of death for persons 5 to 24 years of age.
  - Assaults were the leading cause of death for children ages 0 to 4 years.
  - Gunshot wound (GSW) to the head—mortality risk 75% to 80%. The majority of GSW-related TBIs are self-inflicted.

Geriatric Traumatic Brain Injury

- Risk of TBI increases sharply after age 65.
- TBIs among older adults are more frequently due to falls.
- Severity of TBI and mortality among older adults tend to be higher than that observed in other age groups.

Pediatric Traumatic Brain Injury

- See also section in Chapter 10, Pediatric Rehabilitation, for further details.
- TBI is the leading cause of death in children >1 year of age.
- 10 in every 100,000 children die each year secondary to head injuries.
- Annual incidence of TBI in children is 185 per 100,000.
- Causes of pediatric TBI:
  - Falls (72.8%)
  - Transportation-related (28%)
  - Sports and recreational activities (17%)
  - Assault (7%)
- The most common mechanism of injury for pediatric TBI-related deaths was motor vehicle crashes (CDC Surveillance Report 2018–2019).

PATHOPHYSIOLOGY OF TRAUMATIC BRAIN INJURY

PRIMARY VERSUS SECONDARY INJURY

Primary Injury

- Direct disruption of the brain parenchyma from the shear forces of the impact. It occurs immediately (minutes to hours after the impact) and is not amenable to medical intervention. Primary injury includes the following:
  - 🕮 Contusions: Bruising of the cortical tissue (**Figure 2–1**)
    - 🕮 Diffuse axonal injury (DAI; **Figure 2–2**)
      - Immediate disruption of the axons due to acceleration–deceleration and rotational forces that cause shearing upon impact
      - There is also evidence of a secondary axotomy due to increased axolemmal permeability, calcium influx, and cytoskeletal abnormalities that propagate after the injury.
      - 🕮 Clinically, the coupling of the injury to these structures leads to the picture of white matter punctate petechial hemorrhages characteristic of DAI.

- Impact depolarization:
  - Massive surge in extracellular potassium and glutamate release (excitatory) occurs after severe head injury and leads to excitotoxicity (secondary injury).

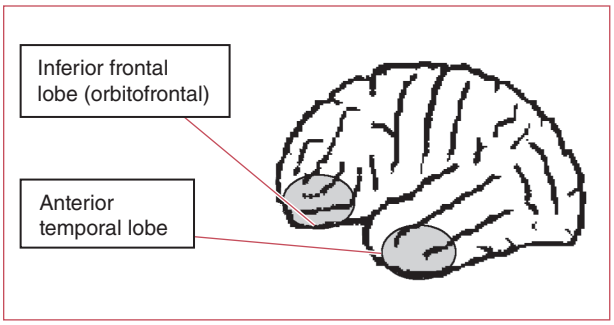

FIGURE 2–1 Location of contusions.

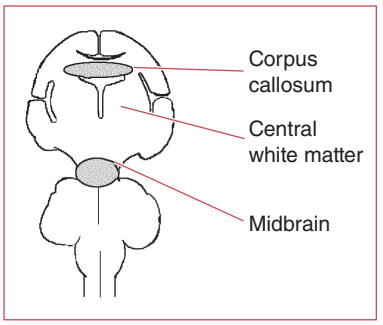

FIGURE 2–2 Common locations of DAI.
DAI, diffuse axonal injury.

Secondary Injury

- Cascade of biochemical, cellular, and molecular events, which include both endogenous cerebral damage as well as extracerebral damage that come with trauma. Mechanisms of secondary injury include the following:
  - Ischemia, excitotoxicity, energy failure, and resultant apoptosis
    - Excitotoxicity is the process by which neuronal damage occurs due to a massive surge in neurotransmitters (also see "Diffuse Injury" section).
  - Secondary cerebral swelling (brain swelling and brain edema)
    - **Brain swelling** occurs early on after acute head injury (within 24 hours) due to an increase in cerebral blood volume (intravascular blood). Identified on CT as collapse of the ventricular system and loss of cerebrospinal fluid (CSF) cisterns around the midbrain.
    - **Brain edema** occurs later after head injury (in comparison to brain swelling) due to an increase in brain volume secondary to increased brain water content → extravascular fluid. There are two types of brain edema:
      1. *Vasogenic edema*:
         - Due to outpouring of protein-rich fluid through damaged vessels
         - Extracellular edema
         - Related to cerebral contusion
      2. *Cytogenic edema*:
         - Found in relation to hypoxic and ischemic brain damage
         - Due to failing of the cells' energy supply system → ↑ cell-wall pumping system → *intracellular* edema in the dying cells
  - Axonal injury
  - Inflammation and regeneration

FOCAL VERSUS DIFFUSE INJURY

Focal Injury

- Localized injury in the brain occurring immediately after the injury and easily visualized by CT or MRI
- Cerebral contusions (see **Figure 2–1**):
  - Occur when the brain impacts the inner table of the skull
  - Occur usually in the inferior frontal lobe and anterior portion of the temporal lobe
- Focal ischemia occurs secondary to vasospasms after a traumatic subarachnoid hemorrhage (SAH) or from physical compression of the arteries.

- 🕮 Focal hemorrhages:
  - **Epidural hematoma (EDH):** Occurs commonly (90%) with a skull fracture in the temporal bone crossing the vascular territory of the middle meningeal artery (60%–90%) or veins (middle meningeal vein, diploic veins, or venous sinus; 10%–40%). Hematoma expansion is slowed by the tight adherence of the dura to the skull.
    - Clinically presents with a lucid interval (50%) prior to rapid deterioration. Biconvex acute hemorrhagic mass seen on head CT (**Figure 2–3**).
  - **Subdural hematoma (SDH):** Occurs in 30% of severe head trauma. They result from shearing of the bridging veins between the pia-arachnoid and the dura. They are usually larger in older adults due to normal age-related cerebral atrophy.
    - High density, crescentic, extracerebral masses seen on head CT (**Figure 2–4**)
    - Acute SDH: Immediately symptomatic lesions
    - Subacute SDH: Those between 3 days and 3 weeks
    - Chronic SDH: Lesions >3 weeks
  - **SAH:** These are closely associated with ruptured cerebral aneurysms and arteriovenous malformations (AVMs) creating blood around the cisterns, although they could also result from leakage from an intraparenchymal hemorrhage and trauma. CT findings demonstrate blood within the cisterns around the brainstem and the subarachnoid space within 24 hours. CT sensitivity decreases to 30% 2 weeks after the initial bleed (**Figure 2–5**).

FIGURE 2–3 Epidural hematoma. *Source:* From Brant WE, Helms CA, eds. *Fundamentals of Diagnostic Radiology*. Lippincott Williams & Wilkins, 2012, with permission.

FIGURE 2–4 Subdural hematoma. *Source:* From Brant WE, Helms CA, eds. *Fundamentals of Diagnostic Radiology*. Lippincott Williams & Wilkins, 2012, with permission.

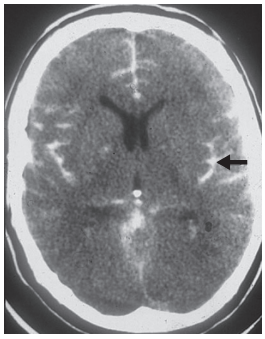

FIGURE 2–5 Subarachnoid hemorrhage (SAH). *Source:* From Giraldo EA. *Subarachnoid hemorrhage (SAH)*. Merck Manual: Professional Version; 2017. https://www.merckmanuals.com/professional/neurologic-disorders/stroke/subarachnoid-hemorrhage-sah.

Diffuse Injury

- Widespread cerebral injury
- DAI is unique to TBI. Its classification is based on severity:
  - *Grade I*: Widespread white matter/axonal damage but no focal abnormalities on imaging
  - *Grade II*: Widespread white matter/axonal damage and focal findings (most common in the corpus callosum)
  - *Grade III*: Damage involving the brainstem
- It is the leading cause of morbidity, including impairments in cognition, behavior, arousal, and coma in TBI. The severity of impairments depends on the magnitude, duration, and direction of angular acceleration of the initial impact.
- 🕮 It is initiated at the time of the injury by axonal shearing from acceleration–deceleration rotational forces, followed by pathophysiologic changes that persist long after the injury. DAI is the most common cause of unconsciousness during and following the first 24 hours of injury.
- Damage is seen most often in the corpus callosum and other midline structures (see **Figure 2–2**): The parasagittal white matter, the interventricular septum, the walls of the third ventricle, and the brainstem (midbrain and pons).

- Pathophysiology:
  - Excitotoxicity: After impact, release of excitotoxic neurotransmitters (glutamate) causes calcium influx and a series of events (oxygen-free radical release, lipid peroxidation, mitochondrial failure, and DNA damage) that ultimately lead to nerve cell death.
  - Hypoxia occurs.
  - Apoptosis: Programmed cell death defined by cell shrinkage, nuclear condensation, and internucleosomal DNA fragmentation with dissolution of the cell membrane. It has both intracellular (cytochrome C, apoptosis-inducing factor [AIF]) and extracellular (tumor necrosis factor [TNF]) triggers.
- Imaging:
  - MRI is more sensitive than CT in revealing DAI, but because axonal injury occasionally has a delayed onset and may or may not be accompanied by edema, diagnostic imaging may not always be reliable.
  - There are now functional MRI studies that can further elucidate dysfunction more clearly than static imaging.

PENETRATING HEAD INJURIES

Missile/Fragments

- Deficits are focal and correspond to the area of injury caused by a bullet/fragment, stab wounds, motor vehicle injury, or occupational injury (e.g., nail).
- If the brain is penetrated at the lower levels of the brainstem, death is instantaneous from respiratory and cardiac arrest. 80% of patients with through-and-through injuries die at once or within a few minutes.
- Mortality rate of patients who are initially comatose from a GSW to the head is 88%, more than two times the mortality rate of closed head injury (CHI).
- Focal or focal and generalized seizures occur in the early phase of the injury in 15% to 20% of cases.
  - Risk of long-term posttraumatic epilepsy (PTE) is higher in penetrating head injuries compared with nonpenetrating injuries.

ANOXIC BRAIN INJURY

- Categorized as nontraumatic brain injury (non-TBI), generally found in metabolically vulnerable areas of the brain, including the parasagittal convexity of the cortex, parieto-occipital cortex, hippocampus, thalamus, putamen, and cerebellum (Fitzgerald et al., 2010).
- Causes include cardiopulmonary arrest, asphyxia, severe anemia, and carbon monoxide poisoning (Fitzgerald et al., 2010).
- Prognosis for recovery is substantially worse for patients with non-TBI compared with TBI (Multi-Society Task Force on PVS, 1994).
- Imaging:
  - MRI is the most sensitive modality for revealing expected diffuse cortical changes. Restricted diffusion in metabolically vulnerable areas can be seen on diffusion-weighted MRI (DW-MRI) in the first 3 to 5 days postinjury (Mason Sharma et al., 2023).
- Treatment:
  - Integral components of acute management include timely initiation of hypothermia, hemodynamic maintenance, treatment of seizures, and supportive care. Seizure prophylaxis is not routinely instituted in cases of anoxic brain injury (Ramiro & Kumar, 2015).

RECOVERY MECHANISMS

🕮 Plasticity

- Brain plasticity represents the capability of the damaged brain to "repair" itself by means of morphologic and physiologic responses.
- Plasticity is influenced by the environment, complexity of stimulation, repetition of tasks, and motivation.

- It occurs via two mechanisms.
  1. **Neuronal regeneration/neuronal (collateral) sprouting**
     - Intact axons establish synaptic connections through dendritic and axonal sprouting in areas where damage has occurred.
     - May enhance recovery of function, may contribute to unwanted symptoms, or may be neutral (with no increase or decrease of function)
     - Thought to occur weeks to months postinjury
  2. **Functional reorganization/unmasking neural reorganization**
     - Healthy neural structures not formerly used for a given purpose are developed (or reassigned) to do functions formerly subserved by the lesioned area.

> Brain plasticity → Remember "PUN"
> **P**lasticity = **U**nmasking + **N**euronal sprouting

Synaptic Alterations

- Include diaschisis and increased sensitivity to neurotransmitter levels

🕮 DIASCHISIS (FIGURE 2–6)

- Mechanism to explain spontaneous return of function
- Lesions/damage to one region of the central nervous system (CNS) can produce altered function in other areas of the brain (at a distance from the original site of injury) that were not severed if there is a connection between the two sites (through fiber tracts). Function is lost in both injured and in morphologically intact brain tissue.
- There is some initial loss of function secondary to depression of areas of the brain connected to the primary injury site, and resolution of this functional deafferentation parallels recovery of the focal lesion (Feeney, 1991).

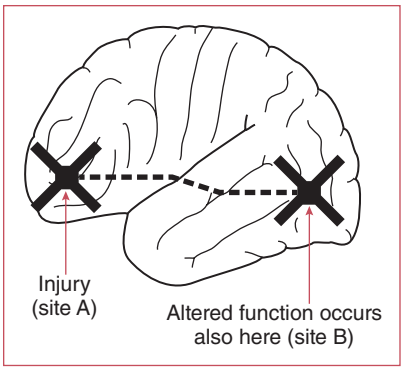

FIGURE 2–6 Example of diaschisis: Injury to site A will produce inhibition of function at site B, which was not severed by the initial injury and is distant from the original site of injury (site A). Recovery of functions controlled by site B will parallel recovery of site A.

Functional Substitution/Behavioral Substitution

- Techniques/new strategies are learned to compensate for deficits and to achieve a particular task.

Redundancy

- Recovery of function based on activity of uninjured brain areas (latent areas) that normally would contribute to that function (and are capable of subserving that function)

Vicariation

- Functions taken over by brain areas not originally managing that function. These areas alter their properties in order to subserve that function.

DISORDERS OF CONSCIOUSNESS

LOCATION OF CONTROL OF CONSCIOUSNESS

🕮 Consciousness

- Consciousness is a function of the ascending reticular activating system (RAS) and the cerebral cortex.

- 🕮 The RAS is composed of cell bodies in the central reticular core of the upper brainstem (mainly midbrain) and their projections to widespread areas of the cerebral cortex via both the thalamic and extrathalamic pathways.
- Lesions that interrupt the metabolic or structural integrity of the RAS or enough of the cortical neurons receiving RAS input can cause disorders of consciousness.

DISORDERS OF CONSCIOUSNESS

	COMA	VEGETATIVE STATE	MINIMALLY CONSCIOUS STATE
Awake?	No	Yes	Yes
Aware?	No	No	Yes (inconsistent but reproducible)

Coma

- Lack of wakefulness as evidenced by the lack of sleep–wake cycles on EEG
- Patient's eyes remain closed.
- There is *no* spontaneous purposeful movement or ability to discretely localize noxious stimuli.
- No evidence of language comprehension or expression
- It results from the damage to the RAS in the brainstem or its connections to the thalami or hemispheres.
- It can last 2 to 4 weeks for people who do not emerge.

🕮 Vegetative State/Unresponsive Wakefulness Syndrome (Laureys et al., 2010)

- Characterized by the resumption of the sleep–wake cycle on EEG
  - No awareness of self or environment
  - No perceivable evidence of purposeful behavior
  - Presence of a verbal or auditory startle but no localization or tracking
  - Patient opens eyes (either spontaneously or with noxious stimuli).
- Neuropathology of vegetative state/unresponsive wakefulness syndrome (VS/UWS):
  - Related to diffuse cortical injury
  - Bilateral thalamic lesions are prominent findings in VS/UWS.
  - In 2019, the American Academy of Neurology (AAN) recommended that the term "chronic VS/UWS" apply to patients in VS/UWS beyond 1 year after TBI and 3 months after non-TBI (Giacino et al., 2019).

Chronic VS/UWS	present >3 months after non-TBI **or** present >12 months after TBI in both children and adults

TBI, traumatic brain injury; UWS, unresponsive wakefulness syndrome; VS, vegetative state.

Minimally Conscious State

- Patient shows minimal but definite evidence of self or exhibits environmental awareness.
- Patient shows evidence of inconsistent but reproducible (or sustained) purposeful behaviors.
  - Simple command following
  - Object manipulation
  - Intelligible verbalization
  - Gestural or verbal yes/no responses
- Patient may also show:
  - Visual fixation
  - Smooth pursuit tracking
  - Emotional or motor behaviors that are contingent upon the presence of specific eliciting stimuli (e.g., patient will cry or get agitated [and behavior is reproducible] only after hearing voices of family members but not with voices of hospital staff)

- Often difficult to differentiate from VS/UWS
- Several evaluations may be required to differentiate minimally conscious state (MCS) from VS/UWS.
- Emergence from MCS typically signaled by:
  - Consistent command following
  - Functional object use
  - Reliable use of a communication system
- Prognosis is better for patients in MCS than in VS/UWS.

TREATMENT OF DISORDERS OF CONSCIOUSNESS

- There is no evidence to support that any kind of therapy-based program (e.g., coma stimulation/sensory stimulation program) will induce or accelerate the cessation of coma or VS/UWS.
- Nevertheless, an organized treatment approach to low-functioning patients permits a quantifiable assessment of responses to stimulation and early recognition of changes or improvements in response to therapeutic interventions or through spontaneous recovery.

Management/Therapy Program

- Neuromedical stabilization
- Preventive therapeutic interventions may be implemented:
  - Manage bowel and bladder function.
  - Maintain nutrition.
  - Maintain skin integrity.
  - Control spasticity.
  - Prevent contractures.
- Pharmacologic interventions:
  - Elimination of unnecessary medicines (e.g., benzodiazepines, H2 blockers, dopamine blockers, pain medications, etc.) and selection of agents with fewest adverse effects on cognitive and neurologic recovery
  - Addition of agents to potentially enhance specific cognitive and physical functions
  - In patients emerging out of coma or VS, the recovery process may be (theoretically) hastened through the use of pharmacotherapy.
  - Agents frequently used include:
    - Dopaminergic agents
      - Amantadine—increases EXOGENOUS dopamine; watch for seizures and nephrotoxicity; increases rate of functional recovery for patients in MCS and VS/UWS (Giacino et al., 2012)
      - Bromocriptine—increases ENDOGENOUS dopamine; watch for hypotension
      - Levodopa/carbidopa—increases EXOGENOUS dopamine
      - Methylphenidate—blocks reuptake of dopamine and norepinephrine
      - Modafinil/armodafinil—stimulates dopamine, histamine, serotonin, norepinephrine, and orexin; armodafinil may result in improved wakefulness relative to modafinil (Darwish et al., 2009); use of modafinil in pregnancy during the first trimester increases risk of congenital malformations (Damkier & Broe, 2020)
    - Acetylcholinesterase inhibitors
    - Antidepressants: Tricyclic antidepressants (TCAs), selective serotonin reuptake inhibitors (SSRIs), and selective serotonin and norepinephrine reuptake inhibitors (SNRIs)
    - Zolpidem—$GABA_A$ receptor agonist, rarely leads to paradoxical improved cognition, about 5% incidence of responders (Whyte et al., 2014)
  - Note: The efficacy of pharmacologic therapy to enhance cognitive function has not been proven.
- Sensory stimulation—widely used despite little evidence of efficacy as previously mentioned
  - Sensory stimulation should include all five senses and address one at a time, in specific therapy sessions and/or in the environmental state, and developed in the room.

– Avoid overstimulation (educate family).
– Patient may have adverse responses due to overstimulation, as ↑ confusion or agitation ↑ reflex responses or avoidance reactions, which may interfere with performance.

POSTURING SECONDARY TO HEAD INJURY

DECEREBRATE POSTURING (FIGURE 2–7A)

- This postural pattern was first described by Sherrington, who produced it in cats and monkeys by transecting the brainstem.
- There is extension of the upper and lower extremities (hallmark: Elbows extended).
- Seen with midbrain lesions/compression; also with cerebellar and posterior fossa lesions
- In its fully developed form, it consists of opisthotonus, clenched jaws, and stiff, extended limbs with internal rotation of arms and ankle plantar flexion (Feldman, 1971).

DECORTICATE POSTURING (FIGURE 2–7B)

- Posturing due to lesions at a higher level (than in decerebrate posture)
- Seen in cerebral hemisphere/white matter, internal capsule, and thalamic lesions
- Flexion of the upper limbs (elbows bent) and extension of the lower limbs

Hint:
Remember, de***COR***ticate → "**COR**" = heart = ♥
⇒ Patient brings hands close to the heart by flexing the elbows.

- Arms are in flexion and adduction and leg(s) are extended.

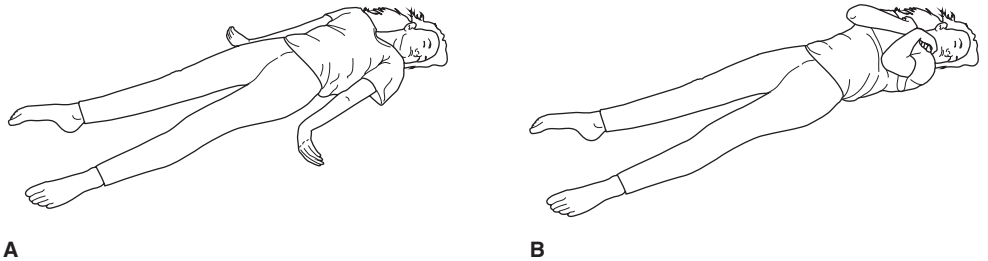

FIGURE 2–7 (A) Decerebrate posture: There is extension of the upper and lower extremities. (B) Decorticate posture: There is flexion of the upper extremities and extension of the lower limbs.

PROGNOSIS AFTER TRAUMATIC BRAIN INJURY: AN EVIDENCE-BASED APPROACH

GLASGOW COMA SCALE (TABLE 2–1)

- 🕮 The GCS is a simple scale for assessing the depth of coma.
- The best GCS score measure in the first 24 hours is used for prognosis. Lower GCS scores are associated with worse outcomes.

TABLE 2–1 Glasgow Coma Scale

SCORE	MOTOR RESPONSE[a]	VERBAL RESPONSE[b]	EYE OPENING[c]
1	None	None	None
2	Decerebrate posturing (extension) to pain	Mutters unintelligible sounds	Opens eyes to pain
3	Decorticate posturing (flexion) to pain	Says inappropriate words	Opens eyes to loud voice (verbal commands)
4	Withdraws limb from painful stimulus	Able to converse—confused	Opens eyes spontaneously
5	Localizes pain/pushes away noxious stimulus (examiner)	Able to converse—alert and oriented	–
6	Obeys verbal commands	–	–

[a]Best motor response—6.
[b]Best verbal response—5.
[c]Best eye opening response—4.
Source: From Teasdale G, Jennett B. Assessment of coma and impaired consciousness. *Lancet*. 1974;304(7872):81–84. doi:10.1016/S0140-6736(74)91639-0, with permission.

- Using the highest GCS score within the first few hours after the injury is preferred, as this reduces the likelihood of using excessively low, very early scores (often before cardiopulmonary resuscitation [CPR]) and confounding factors such as decreased arousal due to use of sedatives or paralytic agents.
- 🕮 Severity of TBI:
  - Severe TBI (coma): GCS score 3 to 8
  - Moderate TBI: GCS score 9 to 12
  - Mild TBI: GCS score 13 to 15
- Total GCS score is obtained from adding the scores of all three categories.
  - Highest score = 15
  - Lowest score = 3
  - GCS score <8: Patient is said to be comatose. The lower the score, the deeper the coma.
- Of the three items in GCS, *best motor response*, particularly 2 weeks postinjury, is the best acute predictor of outcome. The verbal response is the second-best acute predictor of outcome.
- Jennett (1979): Relationship between best GCS score (within the first 24 hours; **Table 2–1**) and outcome as measured on the Glasgow Outcome Scale (GOS; **Table 2–2**):
  - GCS score 3 to 4: Death or VS in 87% of patients
  - GCS score 5 to 7: Death or VS in 53% and moderate or good recovery in 34%
  - GCS score 8 to 10: Moderate or good recovery in 68%
  - GCS score 11: Moderate or good recovery in 87%
  - These data are often missing, as patients arrive paralyzed and intubated.
- Glasgow–Liege Scale: Born et al. (1985) proposed the addition of the brainstem reflex scale to the GCS. Brainstem reflexes in this scale include:
  - Fronto-orbicular reflex (orbicularis oculi): Orbicularis oculi contraction on percussion of the glabella
  - Vertical oculocephalic and horizontal oculocephalic or oculovestibular reflex: "Doll's eye" maneuver (horizontal—moving head forward from side to side; vertical—moving head up and down)
  - Pupillary light reflex
  - Oculocardiac reflex: Bradycardia induced by increasing pressure on the eyeball
  - The Glasgow–Liege Scale has been tested for reliability and prognostic power and has been shown to amplify the information provided by the standard GCS in comatose patients.

TABLE 2–2 Glasgow Outcome Scale

CATEGORY	DESCRIPTION
1. Death	Self-evident criteria
2. VS (alive but unconscious)	Prolonged unconsciousness with no verbalization, no following of commands; absent awareness of self and environment; patient may open eyes; absence of cortical function as judged behaviorally; characterized by the presence of sleep–wake cycles
3. Severe disability (conscious but dependent)	Patient unable to be independent for any 24-hour period by reason of residual mental and/or physical disability
4. Moderate disability (independent but disabled)	Patient with residual deficits that do not prevent independent daily life; patient can travel by public transport and work in a sheltered environment
5. Good recovery (mild to no residual effects)	Return to normal life; there may be minor or no residual deficits

VS, vegetative state.

Coma Duration

- Longer duration of coma is associated with poorer outcomes.
- Metabolic causes of coma have a better prognosis than anoxic causes. The likelihood of good recovery is <5% in those who have suffered SAH or stroke, about 10% in those with hypoxic–ischemic injury, but as high as 25% in those with metabolic causes of coma (Bates, 2001).
- Severe disability is unlikely when coma lasts <2 weeks.
- Good recovery is unlikely when coma lasts >4 weeks.

Posttraumatic Amnesia

- Posttraumatic amnesia (PTA) is one of the most commonly used predictors of outcome.
- A longer duration of PTA is associated with worse outcomes.
- 🕮 Resolution of PTA clinically corresponds to the period when incorporation of ongoing daily events occurs in the working memory.
- *Threshold values*:
  - Severe disability is unlikely when PTA lasts <2 months.
  - Good recovery is unlikely when PTA lasts >3 months.
  - PTA correlates strongly with the length of coma (and with GOS—see later) in patients with DAI but poorly in patients with primarily focal brain injuries (contusions).
- 🕮 **Galveston Orientation and Amnesia Test (GOAT)**—developed by Harvey Levin and colleagues, it is a standard technique for assessing PTA. It is a brief, structured interview that quantifies the patient's orientation and recall of recent events.
  - Assesses orientation to person, place, and time; recall of the circumstances of the hospitalization; and the last preinjury and first postinjury memories
  - 🕮 The end of PTA can be defined as the date when the patient scores 75 or higher in the GOAT for 2 consecutive days. The period of PTA is defined as the number of days beginning at the end of the coma to the time the patient attains the first of two successive GOAT scores ≥75 (Ellenberg et al., 1996).
  - Most important, attempts at more involved neuropsychological assessment usually prove unproductive until the patient consistently obtains scores of 70 or greater. Once a score of 70 is achieved, neuropsychological test data usually are reliable for further rehabilitation and postdismissal planning (DeLisa et al., 2005).
- **Orientation Log (O-Log):** Developed by Jackson, Novack, and Dowler as an alternative assessment of PTA

  - The O-Log was developed because of some of the problems related to the GOAT, including unequal weighting of scored items, difficulty verifying some responses, and a lack of relevancy.
  - It has been shown to have a high correlation to the GOAT (Novack, 2000).
  - The O-Log focuses on orientation to place, time, and circumstance and consists of 10 items scored 0 to 3.
  - Components:
    - Place: Type of place, name, and city
    - Time: Month, date, year, day of week, and time of day
    - Circumstance: Event that made the person require care and the deficits that have resulted.
  - Items are scored 0 to 3: 3 = spontaneous/free recall; 2 = logical cueing; 1 = multiple choice, phonemic cueing; 0 = unable/incorrect/inappropriate.
  - Two consecutive scores of 25 and higher indicate the person is out of PTA.
- Categories of PTA (**Tables 2–3 and 2–4**): Duration of PTA is often used to categorize severity of injury according to the relevant criteria.

TABLE 2–3 Posttraumatic Amnesia Duration and TBI Severity

PTA DURATION	TBI SEVERITY
0–1 days	Mild
>1 day and <7 days	Moderate
>7 days	Severe

PTA, posttraumatic amnesia; TBI, traumatic brain injury.

TABLE 2–4 TBI Recovery Based on PTA Duration

LENGTH OF PTA	LIKELY OUTCOME
1 day or less	Expect quick and full recovery with appropriate management (a few may show persisting disability).
>1 day	Recovery period more prolonged—now a matter of weeks or months.
<1 week	Full recovery possible, for most of these cases, with good management.
1–2 weeks	Recovery a matter of many months. Many patients are left with residual problems even after the recovery process has ended, but one can be reasonably optimistic about functional recovery with good management.
2–4 weeks	Process of recovery is very prolonged—1 year or longer is not unusual. Permanent deficits are likely. There must be increasing pessimism about functional recovery when PTA reaches these lengths.
>4 weeks	Permanent deficits and significant disability likely. It is not just a matter of recovery, but of long-term retraining and management.

PTA, posttraumatic amnesia; TBI, traumatic brain injury.
Source: From Brooks DN, McKinlay WW. *Evidence and Quantification in Head Injury: Seminar Notes*. Unpublished material, 1989, with permission.

Other Indicators of Outcome After Traumatic Brain Injury

- **Age:**
  - Children and young adults tend to have a generally more positive prognosis than older adults. However, young children (<5 years old) and older adults (>65 years old) have greater mortality.
  - Age ≥40 correlates with worse functional outcome when compared with patients <40 (Katz & Alexander, 1994).
- **Rate of early recovery:** Reflected in serial Disability Rating Scale (DRS; see **Table 2–5**), which has been found to be predictive of final outcome. Also see JFK Coma Recovery Scale (JFK-CRS).

- **Pupillary reaction to light:**
  - 50% of patients with reactive pupils after TBI achieve moderate disability to good recovery (in DRS scale) versus 4% with nonreactive pupils.
  - Similarly, the presence of conjugate eye movement is a positive prognostic indicator.
- **Time:**
  - Most recovery usually occurs within the first 6 months postinjury.
- **Postcoma use of phenytoin:**
  - Long-term use of phenytoin has been reported to have adverse cognitive effects (neurobehavioral effects in severe TBI patients compared with placebo group).
- **Decorticate posturing** is a more positive prognostic indicator than decerebrate posturing or flaccid muscle tone (Whyte et al., 2005).
- Also see Table 2–5.

TABLE 2–5 Prognosis in Severe Head Injury

PREDICTIVE INDICATOR	POORER PROGNOSIS	BETTER PROGNOSIS
GCS score	<7	>7
CT scan	Large blood clot; massive bihemispheric swelling	Normal
Age	Old age	Youth
Pupillary light reflex	Pupils remain dilated	Pupil contracts
Doll's eye sign	Impaired	Intact
Caloric testing with ice water	Eyes do not deviate	Eyes deviate to irrigated side
Motor response to noxious stimuli	Decerebrate rigidity	Localizes defensive gestures
SSEP	Deficient	Normal
PTA length	>2 weeks	<2 weeks

GCS, Glasgow Coma Scale; PTA, posttraumatic amnesia; SSEP, somatosensory evoked potential.
Source: From Braddom RL. *Physical Medicine and Rehabilitation*. W.B. Saunders Company; 1996, with permission.

HEAD INJURY PREDICTOR SCALES AND TESTING

Disability Rating Scale (Table 2–6)

- Pros:
  - Widely used scale
  - Documented correlation between acute predictors of outcome and GOS score at 6 months and at 12 months
- Cons:
  - Categories broad with limited sensitivity
  - Not a real indicator of functional abilities
- This is a 30-point scale covering the following eight dimensions (Table 2–6):
  1. Eye opening
  2. Verbalization/communication
  3. Motor responsiveness
  4. Feeding*
  5. Toileting*
  6. Grooming*
  7. Overall level of functioning/dependence
  8. Employability

- The DRS was developed specifically for brain injury.
- It provides a quantitative index of disability.
- It is more sensitive to clinical changes than GOS.
- **Note:* Measuring cognitive skills only in these categories
  - Ten-level global scale that focuses on cognitive recovery and behavior after TBI
  - Gouvier et al., 1987—Levels of Cognitive Function Scale (LCFS; **Table 2–7**) has lower validity and reliability than the DRS (**Table 2–6**).

TABLE 2–6 Disability Rating Scale

1. **Eye Opening** 0 Spontaneous 1 To Speech 2 To Pain 3 None	2. **Communication** 0 Oriented 1 Confused 2 Inappropriate 3 Incomprehensible 4 None	3. **Motor Response** 0 Obeying 1 Localizing 2 Withdrawing 3 Flexing 4 Extending 5 None
4. **Feeding** 0.0 Complete 0.5 1.0 Partial 1.5 2.0 Minimal 2.5 3.0 None	5. **Toileting** 0.0 Complete 0.5 1.0 Partial 1.5 2.0 Minimal 2.5 3.0 None	6. **Grooming** 0.0 Complete 0.5 1.0 Partial 1.5 2.0 Minimal 2.5 3.0 None
7. **Level of Functioning (Physical and Cognitive Disability)** 0.0 Completely independent 0.5 1.0 Dependent in special environment 1.5 2.0 Mildly dependent—limited assistance (nonresident helper) 2.5 3.0 Moderately dependent—moderate assistance (person in home) 3.5 4.0 Markedly dependent—assist all major activities, all times 4.5 5.0 Totally dependent—24-hour nursing care		8. **Employability (as Full-Time Worker, Homemaker, or Student)** 0.0 Not restricted 0.5 1.0 Selected jobs, competitive 1.5 2.0 Sheltered workshop, noncompetitive 2.5 3.0 Not employable

Source: From Rappaport M, Hall KM, Hopkins K, Belleza T, Cope DN. Disability rating scale for severe head trauma: coma to community. *Arch Phys Med Rehabil.* 1982;63:118–123. https://pubmed.ncbi.nlm.nih.gov/7073452/, with permission.

TABLE 2–7 Rancho Los Amigos Levels of Cognitive Function Scale (Rancho Scale or LCFS)

LEVEL	DESCRIPTION
I	No response
II	Generalized response to stimulation
III	Localized response to stimuli
IV	Confused and agitated behavior
V	Confused with inappropriate behavior (nonagitated)
VI	Confused but appropriate behavior
VII	Automatic and appropriate behavior
VIII	Purposeful and appropriate behavior
IX	Purposeful and appropriate (standby assistance on request)
X	Purposeful and appropriate (modified independent)

Source: From Rancho Los Amigos Level of Cognitive Function Scale. National Rehabilitation Center, with permission.

The FIM® Instrument

- Ordinal scale with 18 items and seven levels to assess physical and cognitive function (Table 2–8)
- Documented validity and reliability

TABLE 2–8 The FIM® Instrument

MOTOR ITEMS	COGNITION ITEMS
• Self-care – [a]Eating – [a]Grooming – [a]Bathing – [a]Dressing upper body – [a]Dressing lower body – [a]Toileting – Swallowing	• Communication – [a]Comprehension – [a]Expression – Reading – Writing – Speech intelligibility
• Sphincter control – [a]Bladder management – [a]Bowel management	• Psychosocial adjustment – [a]Social interaction – Emotional status – Adjustment to limitations – Employability
• Mobility – [a]Bed, chair, wheelchair transfer – [a]Toilet transfer – [a]Tub, shower transfer – Car transfer – [a]Walking/wheelchair locomotion – [a]Stairs – Community access	• Cognitive function – [a]Problem-solving – [a]Memory – Orientation – Attention – Safety judgment
Level of Function Scoring	
Independent: • 7 Complete independence (timely, safely) • 6 Modified independence (device)	
Modified Dependence: • 5 Supervision (subject = 100%) • 4 Minimal assistance (subject = 75%+) • 3 Moderate assistance (subject = 50%+)	
Complete Dependence: • 2 Maximal assistance (subject = 25%+) • 1 Total assistance (subject = less than 25%)	

[a]Identifies the 18 items used to assess physical and cognitive function.
Source: From Functional Independence Measure. (Guide for the Uniform Data Set for Medical Rehabilitation, 1996.) FIM is a trademark of Netsmart Technologies, Inc. Reprinted with permission.

Revised JFK Coma Recovery Scale

- Theorizes hierarchical responses (from generalized to cognitively mediated) for 25 items in six areas: Auditory, visual, motor, oromotor/verbal, communication, and arousal.
- Giacino et al. (1991) found that changes in CRS scores showed stronger correlations with outcome (as measured by the DRS) than initial, one-time scores. The change as a percentage of total score was greater for the CRS than for the GCS or DRS (Horn & Zasler, 1996).
- A revised restructured version was published in 2004 (Giacino et al., 2004).

🕮 Neuropsychological Testing

- Prior to the development of the CT scan, neuropsychological assessment was utilized to determine the presence, location, and type of brain lesion a patient had.
- The *Halstead–Reitan Neuropsychological Battery* (HRNB) is used as a diagnostic test for all kinds of brain damage. The original battery was initially designed to assess frontal lobe disorders by Halstead (1947) and subsequently used by Reitan (1970–1974), who added some tests and recommended its use as a diagnostic test for all kinds of brain damage.
  - Most examiners administer this battery in conjunction with the Wechsler Adult Intelligence Scale–Revised (WAIS–R) and Wechsler Memory Scale (WMS) or the Minnesota Multiphasic Personality Inventory (MMPI).
- *WAIS–R*: 11 subtests (six determine verbal IQ and five determine performance IQ); WAIS–R is the most frequently used measure of general IQ. (The revised version of the WAIS–R is known as WAIS–III.)
  - WAIS–III components:
    - Verbal IQ = vocabulary, similarities, arithmetic, digit span, information, comprehension, letter–number sequencing
    - Performance IQ = picture completion, digit–symbol coding, block design, matrix reasoning, picture arrangement, object assembly
- *MMPI*: Consists of 550 true/false questions that yield information about aspects of personality. It is the most widely and thoroughly researched objective measure of personality.
- *Token test*: Correlates with scores on tests of auditory comprehension and language production. Comprised of 20 tokens (circles/squares, big/little, five colors), laid out horizontally. Increasingly complex oral commands, for example, "touch the white circle" → "before touching the yellow circle, pick up the red square." Has 90% sensitivity in discriminating people with aphasias from normals (Lezak, 1995).

Other Indicators of Outcome After Traumatic Brain Injury

AGE

- Older age is associated with worse outcomes.
- Good recovery is less likely in patients older than 65 years.

NEUROIMAGING

- Certain features (depth of injury, bihemispheric lesions) are associated with worse outcomes.
- Good recovery is unlikely when bilateral hemispheric lesions are found on early MRI.

MEDICAL MANAGEMENT OF TRAUMATIC BRAIN INJURY

INITIAL MANAGEMENT

Resuscitation (ABCs)

- Airway management:
  - Endotracheal intubation to maintain oxygenation and airway protection is indicated in patients with GCS <9.
  - Rapid sequence intubation (RSI) with sedating/paralytic agents (succinylcholine, thiopental, fentanyl) could be used in emergent situations. Careful attention should be given to the cervical spine (avoiding neck manipulation/hard cervical collars) in patients with severe TBI or those with a high index of suspicion for spine trauma.
- Oxygenation (breathing):
  - PO_2 should be kept between 100 and 120 mmHg.
  - PCO_2 should be kept between 28 and 32 mmHg.

- Circulation:
  - Keep central perfusion pressure (CPP) >60 mmHg.
  - Remember: CPP = mean arterial pressure (MAP) – intracranial pressure (ICP).
  - Use hypertonic solutions, osmotic diuretics, and crystalloids to decrease ICP.

Laboratory Studies

- Arterial blood gases (ABGs) can determine oxygenation and perfusion.
- Example: Metabolic acidosis is indicative of poor tissue perfusion with good oxygenation.
- Hemoglobin and hematocrit levels are used to monitor blood loss.
- Metabolic panel is used to monitor for hypokalemia and hyperglycemia, both of which can be seen in hyperadrenergic states.
- ETOH and drug screens can provide useful data.

Imaging Studies

- **CT scan:**
  - Has reduced the morbidity and mortality as well as the need for surgical exploration in TBI
  - Early head CT mandatory in GCS <13
  - Should be carried out as soon as the patient has good oxygenation and is hemodynamically stable
  - CT is preferable early on, as it can quickly demonstrate hemorrhagic lesions. However, it is poor in identifying nonhemorrhagic lesions, making correlation with eventual neurologic outcomes poor.
  - Does not define the functional or metabolic status of the brain or provide information on blood flow
- **Brain MRI:**
  - Can define nonhemorrhagic lesions of the brain (cortical contusions, subcortical gray matter injury, and brainstem lesions)
  - Takes much longer (45 vs. 5 minutes) than a head CT and does not really affect early decision-making
  - May come into play later in the management of TBI in terms of defining the lesions and extent of injury as well as guiding treatment options

Serial Neurologic Examinations (Glasgow Coma Scale Scores)

- Changes in pupillary responses in the unresponsive patient can be indicative of a specific pathology or worsening status.
- Anisocoria may indicate impending transtentorial herniation to the ipsilateral side.
- Bilateral fixed dilated pupils indicate a poor outcome.
- Gaze deviations point toward the site of the lesion.
- Rhythmic eye movements might indicate a seizure.
- Loss of corneal and oculocephalic reflexes is indicative of a severe TBI.

Respiratory pattern: Abnormal respiratory patterns may indicate increasing ICP and herniation. Patients initially present with tachypnea and hyperventilation followed by irregular breathing patterns (apnea), bradycardia, and the Cheyne–Stokes pattern of breathing.

Sedation

- *Benzodiazepines* are gamma-aminobutyric acid (GABA) nergic agents often used in controlling agitation and anxiety. They have minimal hemodynamic effects, and lorazepam does not diminish the respiratory drive. They have no direct effect on decreasing ICP, but long-term use can delay neurologic recovery.
- *Barbiturates* can be used to induce a coma and decrease the metabolic requirements of the brain. Pentobarbital can also control ICP.

- *Propofol infusion* can provide the same CNS depression as the barbiturates, and its short half-life can allow for frequent neurologic examinations. It can produce dose-dependent hemodynamic and respiratory depression and should be used only in hemodynamically stable patients. Believed to manifest its effect through the $GABA_A$ system. It can be used for sedation in patients who are intubated in the ICU. It has a relatively favorable side effect profile with a rapid return to consciousness after the infusion is discontinued.

🕮 Evaluation of Intracranial Pressure

- In a normal adult, reclining with the head and the trunk elevated to 45 degrees, normal ICP is between 2 and 5 mmHg.
  - ICP levels up to 15 mmHg are considered harmless.
- **Elevated ICP:** Defined as ICP >20 mmHg for more than 5 minutes
  - Common after severe TBI (53% reported in a recent series)
  - A unilateral mass lesion causes distortion of the brain, a reduction of the CSF volume, ↑ ICP in the closed skull → formation of internal hernias (including tentorial/uncal herniation—see later).
    - 75% of patients with severe TBI die due to mass effect on tissue, shift, the development of internal hernias, and secondary damage to the CNS.
  - If unchecked, ↑ ICP may cause death mainly because of deformation of tissue, brain shifts, herniation, and cerebral ischemia.
  - When ICP >40 mmHg, there is neurologic dysfunction and impairment of the brain's electrical activity.
  - An ICP >60 mmHg is invariably fatal. Pressures of 20 to 40 mmHg are associated with increased morbidity.
  - Fever, hyperglycemia, hyponatremia, and seizures can worsen cerebral edema by ↑ ICP.
- **Cerebral blood perfusion pressure (CPP)** is reduced when ICP is increased.
  - It is more important to maintain an adequate CPP than control only the ICP.
  - CPP is calculated by subtracting ICP from MAP: CPP = MAP – ICP.
  - CPP should remain >60 mmHg to ensure cerebral blood flow.

Indications for Continuous Monitoring of ICP and for Mechanical Ventilation

- Patient in coma (GCS <8) and with CT findings of ↑ ICP (absence of third ventricle and CSF cisterns)
- Deep coma (GCS <6) without hematoma
- Severe chest and facial injuries **and** moderate/severe head injury (GCS <12)
- After evacuation of intracranial hemorrhage if patient in coma (GCS <8) beforehand

CSF, cerebrospinal fluid; GCS, Glasgow Coma Scale; ICP, intracranial pressure.

🕮 FACTORS THAT MAY INCREASE INTRACRANIAL PRESSURE (Ng et al., 2004; Roth et al., 2014)

- Prone and supine body positioning
- Loud noise
- Vigorous physical therapy (PT)
- Chest PT
- Suctioning
- Elevated blood pressure

METHODS USED TO MONITOR INTRACRANIAL PRESSURE

- Papilledema: Rare in the acute stage after brain injury despite the fact that ↑ ICP is frequent
  - Usually occurs bilaterally

  - May indicate presence of intracranial mass lesion
  - Develops within 12 to 24 hours in cases of brain trauma and hemorrhage, but, if pronounced, it usually signifies brain tumor or abscess, that is, a lesion of longer duration
- CT scan (see earlier)—if CT scan equivocal, cisternography may be done.
- Lumbar puncture (LP) if no papilledema (must rule out mass lesion first)
  - LP carries a certain risk of causing fatal shift of brain tissue (i.e., herniation) in the setting of ↑ ICP.
- Intraventricular ICP monitoring (see Intracranial Pressure Management section) in severe TBI

INTRACRANIAL PRESSURE MANAGEMENT

- Elevate head of bed 30 degrees.
- The use of intraventricular ICP catheters to monitor ICPs in severe injuries or those with abnormal CT scans is recommended.
- 🕮 The use of steroids is not recommended. The CRASH trial showed a significant increase in 2-week mortality in those given systemic steroids during the acute phase.
- Barbiturates and decompressive craniotomies decrease ICPs but do not affect mortality and morbidity.
- Osmotic agent, diuretics, and hypertonic solutions are not treatment standards but may have indications for use in increased ICPs.
- A CPP >60 mmHg in adults is recommended. A CPP <50 mmHg is associated with decreased brain tissue oxygenation and increased morbidity and mortality.
- Intubation and hyperventilation: Reduction of $PaCO_2$ through hyperventilation is the most rapid means of lowering ICP. However, although transiently effective in lowering ICP, it should only be used in acute neurologic deterioration. Optimal $PaCO_2$ is approximately 30 mmHg.
- Neurosurgical decompression
- Hypothermia may be used to ↓ ICP and it may protect brain tissue by lowering cerebral metabolism. Marion et al. (1997)—treatment with hypothermia for 24 hours in severe TBI patients (GCS 5–7) associated with improved outcome.

SURGICAL MANAGEMENT IN TRAUMATIC BRAIN INJURY

Indications

- Penetrating injury
- Compound depressed skull fracture
- Epidural hematoma
- Subdural hematoma
- Focal contusion or intracerebral hemorrhage

Emergency Craniotomy

- Definitive care for significant traumatic epidural and SDHs
- 40% of patients with a GCS <9 will have intracranial hematomas. Comatose patients who underwent surgery within 4 hours had better outcomes.
- Performed mainly to remove the source of mass effect or increasing ICP or to clean and debride contaminated open wounds in penetrating trauma

Emergency Burr Hole

- Indicated for a rapidly deteriorating patient who exhibits signs of impending transtentorial herniation caused by an expanding epidural or SDH

Temporal Lobe—Tentorial (Uncal) Herniation (Figure 2–8)

- Uncal herniation results when the medial part of the temporal lobe (uncus and parahippocampal gyrus) is displaced over the edge of the ipsilateral tentorium so as to compress the third cranial nerve (CN), midbrain, cerebral cortex, and subthalamus.
- Occurs as a result of increased supratentorial pressure. It is commonly associated with hematoma (subdural or epidural) secondary to trauma or to a brain tumor.
- Uncal herniation of the medial temporal lobe produces:
  1. Stretching of CN III (oculomotor nerve), which causes ipsilateral pupillary dilation and may lead to complete ipsilateral CN III palsy (fixed pupil dilation, ptosis, and, later, ophthalmoplegia)
  2. Ipsilateral hemiparesis due to pressure on the corticospinal tract located in the contralateral crus cerebri
  3. Contralateral hemiparesis due to pressure (from edema or mass effect) on the precentral motor cortex or the internal capsule
  4. In an uncal herniation, reduced consciousness and bilateral motor signs appear relatively late. Central hyperventilation may also occur late in uncal herniation.

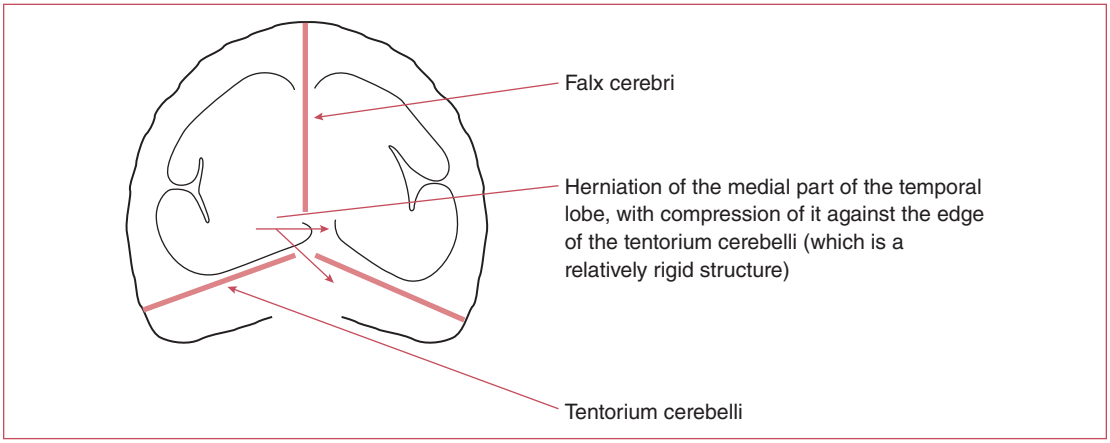

FIGURE 2–8 Temporal lobe—tentorial (uncal) herniation.

MEDICAL AND NEUROLOGIC COMPLICATIONS AFTER TRAUMATIC BRAIN INJURY

POSTTRAUMATIC SEIZURES AND POSTTRAUMATIC EPILEPSY

- Seizure: Clinical manifestation presumed to result from an abnormal and excessive discharge of a set of neurons in the brain
- Posttraumatic seizures (PTS): Seizures occurring within the first 7 days after TBI
- Epilepsy: In a nontraumatic setting, it is a condition characterized by recurrent (at least two) seizures unprovoked by any immediate identified cause that are 24 hours apart.
- Posttraumatic epilepsy (PTE): Recurrent PTS occurring at least a week after TBI and are not attributable to any other etiology than TBI
- PTS are classified as:
  - *Partial seizure* (simple partial if preserved consciousness; complex partial if consciousness is impaired)
  - *Generalized seizure* (grand mal or tonic–clonic)
  - Majority of PTS are simple partial.

- Seizures are also classified by onset.
  - Immediate PTS: Occurs within the first 24 hours postinjury
  - Early PTS: Occurs within the first week (24 hours to 7 days)
  - Late PTS: Occurs after the first week
- **Incidence:**
  - Varies greatly according to the severity of the injury, the time since the injury, and the presence of risk factors (see the text that follows)
  - 5% of hospitalized TBI patients (CHI) have late PTS.
  - 4% to 5% of hospitalized TBI patients have one or more seizures in the first week after the injury (early PTS; Rosenthal et al., 1990).
  - 50% to 66% of PTS occur within 1 year; 75% to 80% occur within 2 years.
  - Most PTS occur 1 to 3 months postinjury.
  - 50% of patients with PTS will have only one seizure, and 25% have no more than three episodes.

TBI Severity and Incidence of Posttraumatic Seizures

- A 1998 study evaluated a group of 4,541 patients with TBI (characterized by LOC, PTA, SDH, or skull fracture) who were divided into three categories:
  - Mild TBI: LOC or amnesia <30 minutes
  - Moderate TBI: LOC for 30 minutes to 24 hours or skull fractures
  - Severe TBI: LOC or amnesia >24 hours, SDH, or brain contusions
- Incidence of seizures in these different categories:
  - Mild TBI: 1.5%
  - Moderate TBI: 2.9%
  - Severe TBI: 17%
  - Overall incidence (all patients): 3.1%

LOC, loss of consciousness; PTA, posttraumatic amnesia; SDH, subdural hematoma; TBI, traumatic brain injury.
Source: From Annegers JF, Hauser WA, Coan SP, Rocca WA. A population-based study of seizures after traumatic brain injuries. *N Engl J Med*. 1998;338(1):20–24. doi:10.1056/NEJM199801013380104, with permission.

🕮 Risk Factors Associated With Late Posttraumatic Seizures (Figure 2–9)

- Penetrating head injury: 33% to 50%
- Intracranial hematoma: 25% to 30%
- Early PTS (>24 hours to 7 days): 25%
- Depressed skull fracture: 3% to 70%
- Prolonged coma or PTA (>24 hours): 35%
- Other risk factors:
  - Dural tearing
  - Presence of foreign bodies
  - Focal signs such as aphasia and hemiplegia
  - Age
  - Alcohol abuse
  - Use of TCAs
- A 2003 prospective, multicenter study (Englander et al., 2003) analyzed risk factors for late PTS:
  - 66% Bilateral parietal contusions
  - 63% Dural penetration
  - 36% Intracranial operation
  - 33% Multiple subcortical contusions
  - 28% SDH, status post evacuation
  - 25.8% >5 mm midline shift

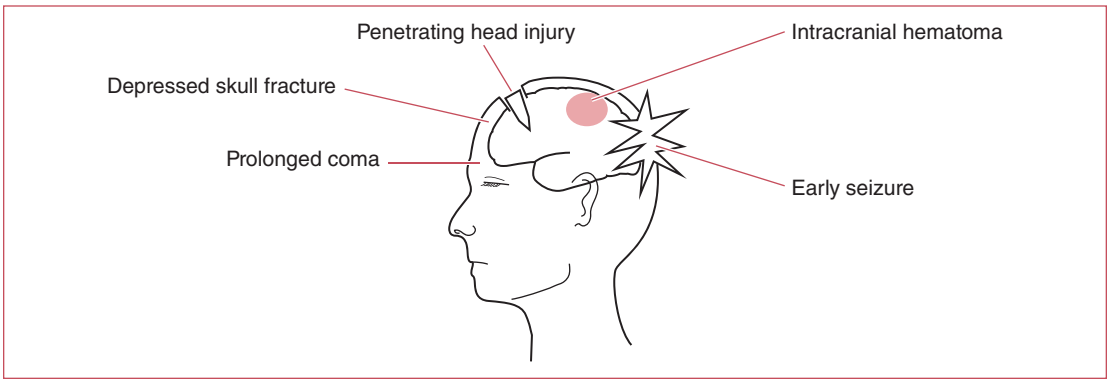

FIGURE 2–9 Risk factors associated with posttraumatic seizures.

Diagnosis of Posttraumatic Epilepsy

- Clinical exam/findings
- EEG (standard, sleep-deprived, 24-hour)
- Prolactin level: ↑ prolactin level confirms true seizure activity, but normal level does not rule out seizure activity. Prolactin level should be drawn within 20 minutes (Chen et al., 2005).

Seizure Prophylaxis

- Greater risk of development of PTS in the first 2 years postinjury
- Prophylactic use of antiepileptic drugs (AEDs) has not been proven effective in prospective, randomized controlled studies.
- Phenytoin and valproic acid have been proven effective only during the first week postinjury at decreasing the incidence of early PTS. No benefit seen with prophylaxis for more than 1 week.
- There is also no proof of change in overall outcome with prophylactic use of phenytoin (Temkin et al., 1990) and evidence it may delay recovery.

Treatment of Posttraumatic Seizure (Chang & Lowenstein, 2003): (Table 2–9)

- Seizure prophylaxis should be initiated immediately and continued for 7 days.
- Immediate PTS: Occurring <24 hours post injury; no AED initiation indicated.
- Early PTS: Occurring within 24 hours and 7 days postinjury; seizure prophylaxis prevents early PTS and lowers risk of seizure recurrence. AED would be continued past 7 days; however, optimal duration is unclear.
- Late PTS: Occurring after 7 days postinjury; seizure prophylaxis does not prevent late PTS; high risk of seizure recurrence; initiate long-term AED.
- 🕮 Levetiracetam (Keppra), carbamazepine, and valproic acid are generally accepted as the drugs of choice for the treatment of PTS. Levetiracetam is now commonly used as a first-line treatment for early PTS prophylaxis due to its favorable side effect profile and comparable efficacy relative to phenytoin, which traditionally was the drug of choice (Jones et al., 2008; Xu et al., 2016; Zafar et al., 2012).
- 🕮 Therapeutic anticonvulsant medications are usually started once late seizures occur. Haltiner et al. (1997) showed that after the first episode of late PTS, the probability of recurrent late PTS is up to 86% within 2 years.
- 🕮 In the TBI population, carbamazepine (partial seizures) and valproic acid (generalized seizures) are often preferred to medications that are more sedating or associated with cognitive impairment (such as phenobarbital and phenytoin). Their superiority over phenytoin has been debated, but the differences among these three agents are probably minimal; carbamazepine may be as sedating as phenytoin (Yablon et al., 1998).
- Important to remember that all anticonvulsants may cause some degree of sedation and cognitive deficits (usually psychomotor slowing).

TABLE 2–9 Antiepileptic Drugs: Uses and Adverse Reactions

MEDICATION	USES	ADVERSE REACTIONS
Carbamazepine	• Partial seizures • Tonic–clonic; generalized seizures • Stabilization of agitation and psychotic behavior • Bipolar affective disorder • Neuralgia	• Acute: Stupor or coma, hyperirritability, convulsions, respiratory depression, Stevens–Johnson syndrome • Chronic: Drowsiness, vertigo, ataxia, diplopia, blurred vision, nausea, vomiting, aplastic anemia, agranulocytosis, hypersensitivity reactions (dermatitis, eosinophilia, splenomegaly, lymphadenopathy), transient mild leukopenia, transient thrombocytopenia, water retention with decreased serum osmolality and sodium, transient elevation of hepatic enzymes
Gabapentin	• Partial seizures	• Somnolence, dizziness, ataxia, fatigue
Levetiracetam	• Partial seizures • Myoclonic seizures • Commonly used as first-line treatment for early PTS prophylaxis	• Acute: Hyperirritability, depression • Brivaracetam (levetiracetam analog) has similar efficacy to levetiracetam; however, behavioral adverse effects are less frequent and severe • Chronic: Drowsiness, vertigo, ataxia, hallucinations, flu-like symptoms, poor coordination, rash, bipolar disorder, suicidality
Phenobarbital	• Partial seizures • Tonic–clonic; generalized seizures	• Sedation, irritability, and hyperactivity in children, agitation, confusion, rash, exfoliative dermatitis, hypothrombinemia with hemorrhage in newborns whose mothers took phenobarbital, megaloblastic anemia, osteomalacia • Nystagmus and ataxia at toxic doses
Phenytoin	• Partial seizures • Tonic–clonic; generalized seizures • Neuralgia	• Intravenous administration: Cardiac arrhythmias, hypotension, CNS depression • Oral administration: Disorders of the cerebellar and vestibular systems (such as nystagmus, ataxia, and vertigo), cerebellar atrophy, blurred vision, mydriasis, diplopia, ophthalmoplegia, behavioral changes (such as hyperactivity, confusion, dullness, drowsiness, and hallucination), increased seizure frequency, gastrointestinal symptoms, gingival hyperplasia, osteomalacia, megaloblastic anemia, hirsutism, transient liver enzyme elevation, decreased antidiuretic hormone secretion leading to hypernatremia, hyperglycemia, glycosuria, hypocalcemia, Stevens–Johnson syndrome, systemic lupus erythematosus, neutropenia, leukopenia, red cell aplasia, agranulocytosis, thrombocytopenia, lymphadenopathy, hypothrombinemia in newborns whose mothers received phenytoin, reactions indicative of drug allergy (skin, bone marrow, and liver function)
Valproic acid	• Partial seizures • Tonic–clonic; generalized seizures • Myoclonic seizures • Absence seizures • Stabilization of agitation and psychotic behavior	• Transient gastrointestinal symptoms such as anorexia, nausea, and vomiting; increased appetite; sedation; ataxia; tremor; rash; alopecia; hepatic enzyme elevation, fulminant hepatitis (rare, but fatal); acute pancreatitis; hyperammonemia
Lamotrigine (Messenheimer et al., 1994; Stahl, 2020)	• Partial seizures • Tonic–clonic; generalized seizures • Bipolar 1 disorder	• Benign rash (approximately 10% of users), ataxia, diplopia, headaches, sedation, insomnia, nausea

(*continued*)

TABLE 2–9 Antiepileptic Drugs: Uses and Adverse Reactions (*continued*)

MEDICATION	USES	ADVERSE REACTIONS
Lacosamide (FDA, 2010; Li et al., 2020)	• Partial seizures	• Headache, dizziness, ataxia, suicidal ideation, cardiac conduction problems, syncope, skin rashes, hepatotoxicity
Topiramate (Stahl, 2020)	• Partial seizures • Tonic–clonic; generalized seizures • Migraine prophylaxis • Weight management	• Sedation, dizziness, ataxia, paresthesia, nystagmus, anorexia, blurred vision, diplopia, impaired concentration

CNS, central nervous system; PTS, posttraumatic seizures.
Source: Adapted from Rosenthal M, Griffith ER, Kreutzer JS, Pentland B, eds. *Rehabilitation of the Adult and Child With Traumatic Brain Injury*. 3rd ed. F. A. Davis; 1999, with permission.

- Phenobarbital is clearly associated with greater cognitive impairment and should not be used as first choice of anticonvulsant therapy in the TBI patient.
- Long-term use of phenytoin has been associated with adverse cognitive effects. Animal and clinical (extrapolated from strokes) studies suggest that phenytoin may impede recovery from brain injury (Dikmen et al., 1991).
- Second-generation anticonvulsants, such as gabapentin and lamotrigine may also be used for treatment of PTS as adjuvant agents (not approved yet for monotherapy). These agents appear to have fewer cognitive side effects.

🕮 Antiepileptic Drug Interactions (Table 2–10)

TABLE 2–10 Antiepileptic Drugs: Common Drug Interactions

MEDICATION	DRUG INTERACTIONS
Carbamazepine	• Increased metabolism of carbamazepine (decreased levels) with phenobarbital, phenytoin, and valproic acid • Enhances metabolism of phenobarbital • Enhances metabolism of primidone into phenobarbital • Reduces concentration and effectiveness of haloperidol • Carbamazepine metabolism inhibited by propoxyphene and erythromycin
Lamotrigine	• When used concurrently with carbamazepine, may increase levels of 10,11-epoxide (an active metabolic of carbamazepine) • Half-life of lamotrigine is reduced to 15 hours when used concurrently with carbamazepine, phenobarbital, or primidone • Reduces valproic acid concentration
Levetiracetam	• When used concurrently with antihistamines, or medications that potentiate GABA, may cause changes in mental status and/or respiratory depression
Phenobarbital	• Increased levels (as much as 40%) of phenobarbital when valproic acid administered concurrently • Phenobarbital levels may be increased when concurrently administering phenytoin • Phenobarbital has a variable reaction with phenytoin levels
Phenytoin	• Phenytoin levels may increase with concurrent use of chloramphenicol, cimetidine, dicumarol, disulfiram, isoniazid, and sulfonamides • Free phenytoin levels may increase with concurrent use of valproic acid and phenylbutazone • Decreased total levels of phenytoin may occur with sulfisoxazole, salicylates, and tolbutamide • Decreased phenytoin levels with concurrent use of carbamazepine • Decreased carbamazepine levels with concurrent use of phenytoin

(*continued*)

TABLE 2–10 Antiepileptic Drugs: Common Drug Interactions (*continued*)

MEDICATION	DRUG INTERACTIONS
Phenytoin (*cont.*)	• Increased or decreased levels of phenytoin when concurrently administered with phenobarbital • When concurrently used with theophylline, phenytoin levels may be lowered and theophylline metabolized more rapidly • May decrease effectiveness of oral contraceptives • Enhances metabolism of corticosteroids
Valproic acid	• Increases level of phenobarbital • Inhibits metabolism of phenytoin • Rare development of absence status epilepticus associated with concurrent use of clonazepam

GABA, gamma-aminobutyric acid.
Source: Modified from Rosenthal M, Griffith ER, Kreutzer JS, Pentland B, eds. *Rehabilitation of the Adult and Child With Traumatic Brain Injury*. 3rd ed. F. A. Davis; 1999, with permission.

WITHDRAWAL OF ANTIEPILEPTIC DRUGS FOR PATIENTS WITH POSTTRAUMATIC EPILEPSY

- 🕮 It is reasonable to consider withdrawal of AEDs after a 2-year, seizure-free interval.
- Increased risk of recurrence for those with more frequent recurrences, treatment with more than one AED, a history of generalized tonic–clonic seizures, and abnormal or epileptiform discharges on prewithdrawal EEGs

Surgical Treatment of Posttraumatic Epilepsy

- The multicenter Study of Epilepsy Surgery reports that resective surgery significantly reduced seizure recurrence after medial temporal (77% 1-year remission) and neocortical resection (56% 1-year remission). There was also significant improvement in terms of quality of life and social functioning.
- Vagal nerve stimulation (VNS), deep brain stimulation of the thalamic anterior nucleus, and responsive neurostimulation are neuromodulatory therapies that offer significant reductions in seizure frequency for patients with partial epilepsy.
- VNS is approved by the Food and Drug Administration (FDA) for adjunctive treatment of intractable partial seizures in patients over 12 years old.
- Surgery is a very effective treatment for intractable frontal lobe epilepsy (FLE) secondary to encephalomalacias.

PAROXYSMAL AUTONOMIC INSTABILITY AND DYSTONIA

- Hypertension (HTN), tachycardia, hyperthermia, spasticity, and perspiration due to a surge of circulating catecholamines released from direct trauma to the autoregulatory centers. It usually occurs within the first 2 weeks of injury.

Management

- Lipophilic beta-blockers for HTN/tachycardia (i.e., propranolol)
- Dantrolene sodium for malignant hyperthermia
- Nonsteroidal anti-inflammatory drugs (NSAIDs; indomethacin), acetaminophen
- Cooling blankets, nasogastric tube (NGT) lavage
- Dopamine agonists: Amantadine, bromocriptine:
  - Anterior hypothalamus—temperature sensitive
  - Posterior hypothalamus—heat dissipation center

POSTTRAUMATIC HYDROCEPHALUS

- Ventriculomegaly (ventricular dilation) is common after severe TBI (40%–72%).
  - Ventriculomegaly is usually due to cerebral atrophy and focal infarction of brain tissue (*hydrocephalus ex-vacuo*).

- However, true posttraumatic hydrocephalus (PTH) is relatively rare. Incidence is 3.9% to 8%.
- PTH in TBI patients is most often of the communicating or normal-pressure type.
- *PTH symptoms*:
  - Classic triad of incontinence, ataxia/gait disturbance, and dementia of normal pressure hydrocephalus is of little help, especially in severe TBI cases.
  - Initial manifestations of hydrocephalus can be intermittent headache, vomiting, and mental status changes (confusion, drowsiness).
- Head CT and further workup to rule out hydrocephalus should be considered if there is failure to improve or deterioration of cognitive or behavioral function.
  - CT findings: Periventricular lucency, sulcal effacement, and uniform ventricular dilation favor PTH.
- Treatment: LP, shunt placement

CRANIAL NERVE INJURIES IN TRAUMATIC BRAIN INJURY

- Most frequently affected CNs (see the text that follows for more detail):
  - Olfactory nerve (CN I)—most common
  - Facial nerve (CN VII)
  - Audiovestibular/vestibulocochlear nerve (CN VIII)
- CNs affected with intermediate frequency:
  - Optic nerve (CN II)—see the text that follows
  - Oculomotor nerves (CN IV > CN III > CN VI)
- Rarely involved CNs: Trigeminal nerve (CN V) and the lower CNs

🕮 CRANIAL NERVE I (OLFACTORY NERVE)

- CN most often damaged by blunt head trauma due to tearing of the olfactory nerve filaments in or near the cribriform plate through which they traverse
- Symptoms: Anosmia and an apparent loss of taste
- Overall incidence approximately 7% and increases with the TBI severity (19.4% in moderate TBI and 24.5% in severe TBI)
- CN I injury is the most likely CN neuropathy present in mild TBI.
- It is often associated with:
  - CSF rhinorrhea
  - Anosmia: Complete loss of smell
  - Dysosmia: Impaired sense of smell
  - Hyposmia: Partial loss of smell
  - Parosmia: Sensation of smell in the absence of stimulus
  - Cacosmia: Awareness of disagreeable or unusually offensive odor that does not exist; may be part of an aura prior to a seizure onset
- In higher level patients, it could present with decrease in appetite, weight loss, and/or altered feeding pattern.
- Recovery occurs in more than one-third of cases, usually during the first 3 months.

CRANIAL NERVE IV (TROCHLEAR NERVE)

- 🕮 Damage to the trochlear nerve can result in vertical diplopia.

CRANIAL NERVE VII (FACIAL NERVE)

- The facial nerve innervates the following four components:
  - Tactile sensation to the parts of the external ear
  - Taste sensation to the anterior two-thirds of the tongue
  - Muscles of facial expression
  - Salivary and lacrimal glands
- It is especially vulnerable to penetrating or blunt trauma to the head because of its long, tortuous course through the temporal bone.

CRANIAL NERVE VIII (VESTIBULOCOCHLEAR NERVE)

- Damage to the vestibulocochlear nerve results in loss of hearing or in postural vertigo and nystagmus coming on immediately after the trauma.

CRANIAL NERVE II (OPTIC NERVE)

- Partial damage may result in scotomas and a troublesome blurring of vision, or as homonymous hemianopsia.
- If CN II is completely involved or transected, patient will develop complete blindness (pupil dilated, unreactive to direct light but reactive to light stimulus to the opposite eye due to consensual light reflex).

POSTTRAUMATIC AGITATION

- Agitation is a subtype of delirium occurring during PTA and is characterized by excesses of behavior, including aggression, akathisia, disinhibition, and/or emotional lability.
- Occurs as patients become more responsive in early stages of recovery
- Usually lasts 1 to 14 days but can last longer
- Most commonly occurs with frontotemporal lesions, which coordinate arousal, attention, executive control, memory, and limbic behavioral functions
- It is important to clearly identify the problem. The generic word *agitation* is not enough; identify the problem.
- Objective measurement is critical. Posttraumatic agitation can be quantified with the Agitated Behavior Scale (ABS) or Overt Agitation Severity Scale (OASS).
  - 🕮 **ABS:** Designed for serial assessment of agitated patients. Ratings are based on behavioral observations made after an 8-hour nursing shift or therapy treatment session. Consists of 14 items or behaviors rated between 1 (absent) and 4 (present to an extreme). Scoring: Below 21: normal; 22 to 28: mild agitation; 29 to 35: moderate agitation; 35 to 54: severe agitation.
  - **OASS:** Contains 47 observable characteristics of agitation to assess its severity. Behavior subgroups are scored 1 to 4 (mild–severe) and multiplied by their frequency for a composite score.

🕮 First-Line Interventions for Posttraumatic Agitation (Table 2–11)

- Patient should be maintained in a safe, structured, low-stimulus environment, which is frequently adequate to manage short-term behavior problems. Agitation may be controlled with alterations in environment and staff or family behavior.
- Floor beds can eliminate the need for restraints (**Figure 2–10**).
- Use physical restraints only if the patient is a danger to self or others. They should be applied only to a minimal degree and should not be a substitute for a floor bed, 1:1 supervision, or other environmental interventions.
- Environmental modifications should be considered prior to proceeding to pharmacologic management.

Second-Line Interventions for Posttraumatic Agitation: Pharmacotherapy

ANTIPSYCHOTIC AGENTS

- Review of dopamine pathways
  - Mesolimbic: Decreased dopamine, decreased positive symptoms
  - Mesocortical: Decreased dopamine, increased negative symptoms
  - Nigrostriatal: Decreased dopamine, increased movement disorders
  - Tuberoinfundibular: Decreased dopamine, increased prolactin
  - Antipsychotic medications can potentially cause neuroleptic malignant syndrome (fever, leukocytosis, muscle stiffness) → treat with dantrolene and beta-blockers.

TABLE 2–11 Environmental Management of Posttraumatic Agitation

1. **Reduce the level of stimulation in the environment:** – Place patient in quiet, private room. – Remove noxious stimuli if possible—tubes, catheters, restraints, and traction. – Limit unnecessary sounds—TV, radio, and background conversations. – Limit number of visitors. – Staff to behave in a calm and reassuring manner. – Limit number and length of therapy sessions. – Provide therapies in patient room.
2. **Protect patient from harming self or others:** – Place patient in a floor bed with padded side panels (Craig bed). – Assign 1:1 or 1:2 sitter to observe patient and ensure safety. – Avoid taking patient off unit. – Place patient in locked ward.
3. **Reduce patient's cognitive confusion:** – One person speaking to patient at a time. – Maintain staff to work with patient. – Minimize contact with unfamiliar staff. – Communicate with patient briefly and simply, one idea at a time.
4. **Tolerate restlessness when possible:** – Allow patient to thrash about in floor bed. – Allow patient to pace around unit with 1:1 supervision. – Allow confused patient to be verbally inappropriate.

Source: From Braddom RL. *Physical Medicine and Rehabilitation*. W.B. Saunders Company; 1996, with permission.

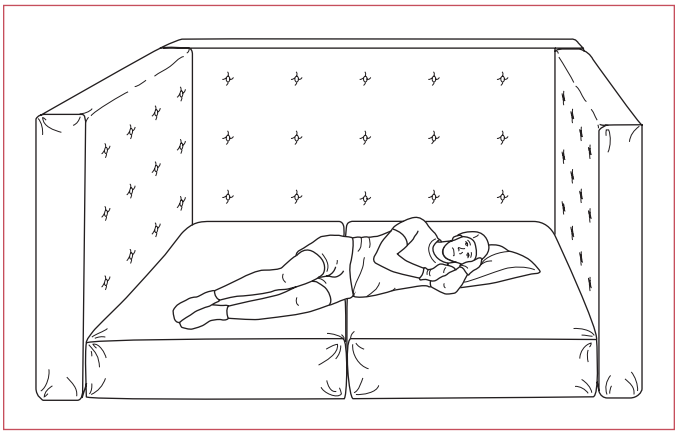

FIGURE 2–10 Agitated, nonambulatory patients often benefit from the use of a floor (Craig) bed. Mattresses can be placed on the floor with 3- to 4-foot padded walls on four sides that allow the patient to roll around. The use of a floor bed with 1:1 supervision and with the use of mitts and a helmet (if necessary) often eliminates the need for restraints.

TYPICAL ANTIPSYCHOTIC AGENTS

- Block D_2-receptors, as well as histaminic, alpha-1-adrenergic, and cholinergic receptors (orthostasis, dry mouth, constipation, and blurry vision). Because acetylcholine and dopamine have a reciprocal relationship in the nigrostriatal pathway, drugs with more anticholinergic properties will increase dopamine in this pathway, lessening extrapyramidal symptoms (EPS).
- Haloperidol has been shown to slow motor recovery in animal models and prolong PTA in humans by causing irreversible dopamine blockade (Feeney et al., 1982). Rapid onset of action.
- Chlorpromazine
- Thiothixene

ATYPICAL ANTIPSYCHOTIC AGENTS

- Less blockage of dopamine D_2-receptors with more serotonin blockade at $5HT_2$-receptor
- Atypicals are less likely to cause motor side effects than typicals (tardive dyskinesia, parkinsonism, dystonia, and akathisia).
- Frequent metabolic adverse effects:
  - Hyperglycemia and development of diabetes
  - Weight gain (more so with clozapine and olanzapine)
  - Hyperlipidemia (more so with clozapine, olanzapine, and quetiapine)
  - Stroke: Only studied with risperidone; demented older adult patients treated with risperidone experienced more transient ischemic attacks (TIAs) and strokes than placebo-treated patients
  - QT prolongation
- Risperidone (Risperdal®):
  - Most "typical" of the atypicals
  - At higher doses, higher incidence of EPS than other atypicals
  - Least anticholinergic; can be stimulating
  - Initial insomnia, agitation, hypotension, which resolves with time
  - Increased prolactin levels
  - While very limited, may have the greatest amount of literature in the TBI population
- Ziprasidone (Geodon®):
  - Most known for QT prolongation; otherwise, favorable side effect profile
  - Least weight gain and risk for diabetes
  - More activating than other antipsychotics at low doses
  - Can be given intramuscularly (IM); therefore, fast onset
- Quetiapine (Seroquel®):
  - Very sedating; therefore, often used for sleep
  - Minimal motor side effects or prolactin elevation
  - Lower likelihood of inducing EPS
  - Dopamine blockaded only with high dosing, at least 400 mg
  - Initial anticholinergic side effects (syncope, hypotension)
- Olanzapine (Zyprexa®):
  - Dose-related EPS, though less than risperidone (above 7.5 mg)
  - Somnolence and gait disturbances common; therefore, best if given at bedtime
  - High rate of metabolic side effects and weight gain
  - Short-acting IM form
- Clozapine (Clozaril®):
  - Serious side effects: Agranulocytosis (monitor white blood cells [WBCs] every 2 weeks), cardiac effects, lowered seizure threshold; intense monitoring required
  - Most anticholinergic activity of all atypicals causing sedation
  - Most weight gain due to antihistaminic properties
  - However, very effective in treating positive symptoms when other treatments have failed
- Aripiprazole (Abilify®):
  - Unique in that it acts as a D_2-antagonist under hyperdopaminergic conditions and D_2-agonist under hypodopaminergic conditions; serotonin agonist at some receptors, antagonist at others
  - Least sedating, fewest EPS, low propensity for metabolic adverse reactions

BENZODIAZEPINES

- Potentially detrimental to patients with stroke and brain injury
  - In cortically injured rats, early daily administration impaired motor recovery and late administration caused transient recurrence of hemiparesis.
  - May cause paradoxical agitation in older adults
  - Amnesic effects may increase confusion in those emerging from PTA.
  - Other side effects include respiratory depression, disinhibition, and impaired coordination.

- If necessary, use midazolam or lorazepam due to short duration of action.
- Good for treatment of spasticity through GABA potentiation
- May have some potential for treatment of mutism in TBI

BETA-BLOCKERS

- 🕮 *Cochrane Review*: Best evidence for efficacy in treating posttraumatic agitation
- No adverse effect on motor recovery but may cause depression and lethargy at higher doses
- Lipophilic agents (propranolol, metoprolol) theoretically most effective:
  - Propranolol can be used up to 520 mg/day: In one study, reduced intensity but not frequency of agitation; significantly reduced number of assaults and attempted assaults in another study.
  - One case study showed metoprolol to be helpful.
- Use is limited by hypotension and bradycardia.
- Also useful for treatment of hyperadrenergic states common in acute TBI

ANTICONVULSANTS (MOOD STABILIZERS)

- Valproic acid (Depakote, Depakene):
  - Various studies have shown it to reduce behavioral outbursts and agitation (two case reports and one case series).
  - Side effects: Sedation, alopecia, tremor, ataxia, gastrointestinal (GI) upset, weight gain
  - Maximum dose limited by hepatotoxicity, thrombocytopenia, and medication toxicity
  - Multiple drug–drug interactions (e.g., lamotrigine, carbamazepine, phenytoin, phenobarbital, rifampin, cimetidine, and aspirin)
  - May have increased metabolism in TBI patients and may require higher doses
- Carbamazepine (Tegretol®), oxcarbazepine (Trileptal®):
  - Can improve irritability, disinhibition, and aggression, though evidence is limited
  - Side effects: Hyponatremia, renal failure, aplastic anemia/agranulocytosis, Stevens–Johnson syndrome, balance disorders, and sedation
  - Inducer of CYP450 3A4
  - Rapid onset
  - Serum levels need to be monitored.
  - May cause some cognitive decline
- Gabapentin (Neurontin®):
  - Helpful in modulating agitation from dementia, but one TBI case study showed increased anxiety and restlessness
- Lamotrigine (Lamictal®):
  - Little weight gain or sedation
  - High rate of benign rashes; serious dermatologic reactions including Stevens–Johnson syndrome have been known to occur
  - Interacts with valproic acid

ANTIDEPRESSANTS

- Metabolites of norepinephrine and serotonin have been found to be reduced in the CSF of agitated anoxic brain injury patients.
- Nortriptyline, amitriptyline, and desipramine have been shown to reduce agitation and aggressive behaviors possibly due to sedative effects; however, not recommended for long-term use due to high fall risk and cognitive impairment (Writer & Schillerstrom, 2009).
- Sertraline was shown in three studies to reduce irritability and aggressive behavior but had no effect in another study.
- Trazodone has been shown to reduce agitation and aggressive behaviors in dementia patients. It also increases duration of deep sleep (Larson & Zollman, 2010).
- Buspirone: Several case studies/series have shown reduced aggressive behaviors.

- Bupropion significantly reduced restlessness in one patient. Bupropion appears to be safe in pregnancy and has not been found to increase rates of major fetal malformation (Chun-Fai-Chan, 2005). Bupropion lowers seizure threshold and should be avoided within 2 weeks post-TBI (Ripley et al., 2018).

LITHIUM

- Improvements in aggressive episodes in several case series/reports
- Significant adverse reactions at high serum levels may limit use (movement disorders, seizures, hypothyroidism, bradycardia, and vomiting).
- Levels must be monitored.
- It may be good for TBI patients whose aggression is related to manic effects and for those whose recurrent irritability is related to cyclic mood disorders.
- Lithium levels will rise if patient becomes dehydrated.

NEUROSTIMULANTS

- Amantadine—unclear, possibly stimulates dopamine; stimulates arousal, memory, and initiation
- Has been shown to reduce agitation in dementia patients and agitation in TBI patients, though other studies showed no difference with amantadine use
- Methylphenidate—blocks reuptake of dopamine and norepinephrine; strengthens neuronal transmission to the amygdala—responsible for learning and emotional memory
  - Had mixed results in behavioral function; may improve anger, but one case report reported increased agitation
  - Use in early pregnancy significantly associated with fetal cardiac malformations
- Dextroamphetamine—blocks reuptake of dopamine and norepinephrine; also a monoamine oxidase inhibitor (MAOI)
- One case study showed positive results.

MEDROXYPROGESTERONE ACETATE (DEPO-PROVERA)

- For aggressive hypersexual behavior—lowers testosterone
- No effect on memory or learning
- Lowers seizure threshold, causes weight gain, increases blood sugar

Management of Pathologic Behaviors in Posttraumatic Agitation (Figure 2–11)

- Identify if this is an emergency issue that requires immediate intervention (severity, potential risk, and acuity).
- Consider possible differential diagnosis:
  - Drug withdrawal
  - Delirium tremens (DTs)
  - Infection
  - Pain
  - Hypoxia
  - Seizure disorder
- Consider environmental issues (see First-Line Interventions for Posttraumatic Agitation section and **Table 2–11**):
  - Low-stimulation environment
  - Reduction of physical discomfort
  - Reduction of lines/direct restraints
  - Reorientation
  - Scheduled toileting program
  - Evaluating and treating sleep–wake cycles

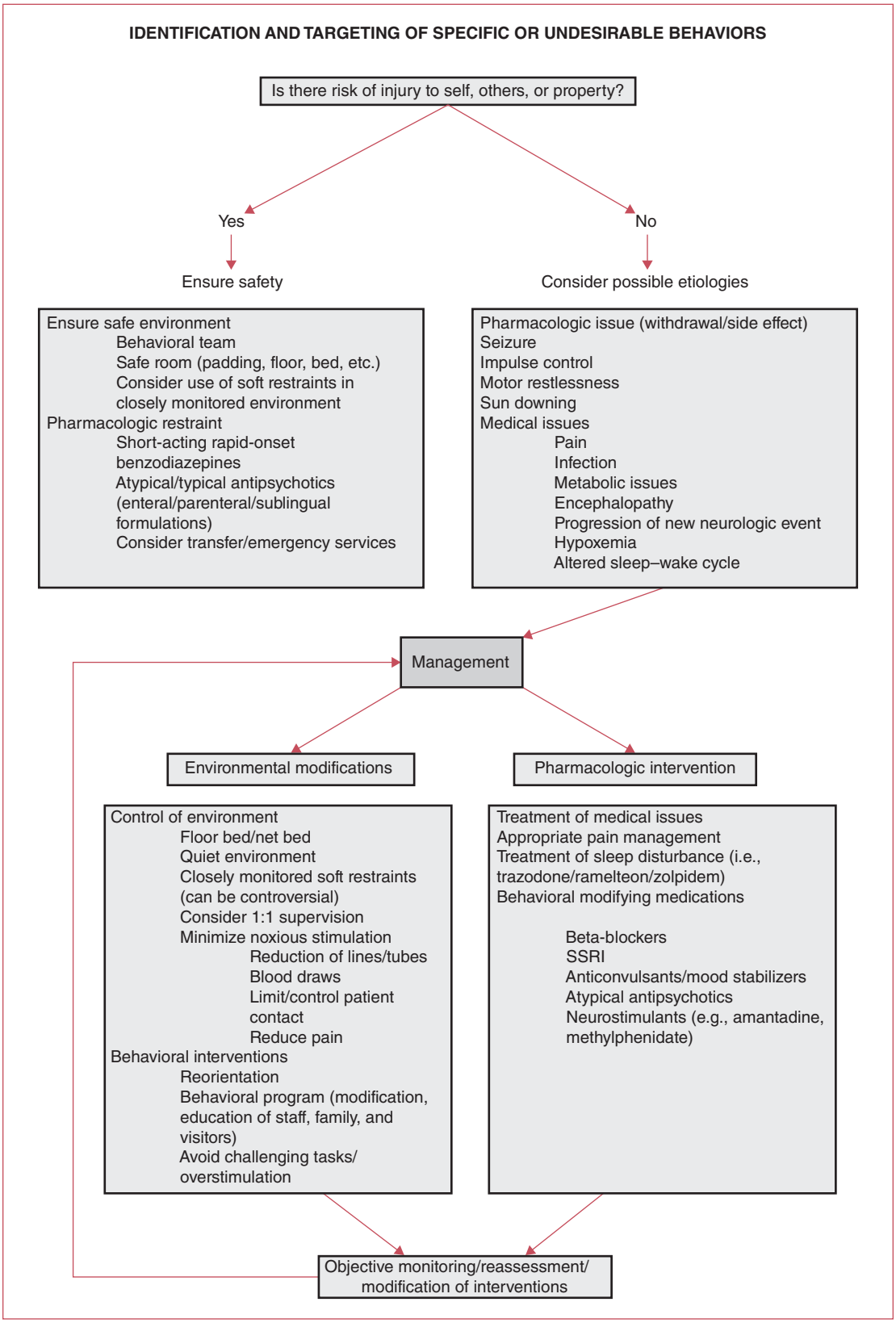

FIGURE 2–11 Agitation management flowsheet.
SSRI, selective serotonin reuptake inhibitor.

- Medication management:
  - Minimize cognitive-impairing medications (benzodiazepines, typical antipsychotics).
  - For immediate effect if there is significant risk of injury to person or property: Atypical antipsychotic
  - Maintenance later with anticonvulsants, beta-blockers, atypical antipsychotics, trazodone, SSRIs, and rarely lithium. For more mild agitation, any of the previously noted maintenance drugs can be used, as well as buspirone. Reassessment with objective measures.

🕮 ALCOHOL WITHDRAWAL

- Studies estimate that 30% and 50% of patients were intoxicated at the time of TBI, with greater estimates for patients injured in MVAs and assaults (Weil et al., 2018).
- Alcohol withdrawal looks like paroxysmal autonomic instability with dystonia syndrome (PAID syndrome) but without dystonia.
- Management involves supportive care; the Clinical Institute Withdrawal Assessment for Alcohol (CIWA) protocol, however, limits benzodiazepine use as it is detrimental to neural recovery.
  - Phenobarbital for alcohol withdrawal as an alternative to benzodiazepines has been studied with shorter hospital stays and lower incidence of invasive mechanical respiration (Tidwell et al., 2018).

🕮 PSEUDO-DEPRESSIVE SYNDROME

- Pseudo-depressive syndrome results from structural brain damage generally to the prefrontal convexity, manifested by neurobehavioral sequelae that overlap with depression, such as apathy, decreased initiative, indifference, perseveration, and inflexibility (Sudarsanan et al., 2007).
- Diagnosis is by neuropsychological testing, which can rule out major depressive disorder and guide treatment (Sudarsanan et al., 2007).
- Treatment includes cognitive behavioral therapy, and serotonergic and dopaminergic agents (Fann et al., 2009; Sudarsanan et al., 2007).

HETEROTOPIC OSSIFICATION

- Heterotopic ossification (HO) is the formation of mature lamellar bone in extraskeletal soft tissue.
- Common in TBI: Incidence of 11% to 76%
  - Incidence of clinically significant cases is 10% to 20%.

Risk Factors

- Prolonged coma (>2 weeks)
- Immobility
- Limb spasticity/↑ tone (in the involved extremity)
- Associated long-bone fracture
- Pressure ulcers
- Edema
- Period of greater risk to develop HO is 3 to 4 months postinjury.

Signs/Symptoms

- Most common: Pain and ↓ range of motion (ROM)
- Also: Local swelling, erythema, warmth in joint, muscle guarding, low-grade fever
- In addition to pain and ↓ ROM, complications of HO include bony ankylosis, peripheral nerve compression, vascular compression, and lymphedema.

- Joints most commonly involved:
  1. 🕮 Hips (most common)
  2. Elbows/shoulders
  3. Knees

Differential Diagnosis

- Deep vein thrombosis (DVT), tumor, septic joint, hematoma, cellulitis, and fracture

DIAGNOSTIC TESTS

SERUM ALKALINE PHOSPHATASE

- Serum alkaline phosphatase (SAP) elevation may be the earliest and least expensive method of detection of HO.
- It has poor specificity (may be elevated for multiple reasons, such as fractures, hepatic dysfunction, etc.).

TRIPLE-PHASE BONE SCAN

- Sensitive method for early detection of HO
- HO can be seen within the first 2 to 4 weeks after injury in Phase I (blood-flow phase) and Phase II (blood-pool phase) of a triple-phase bone scan, and in Phase III (static phase/delayed images) in 4 to 8 weeks, with normalization by 7 to 12 months.

PLAIN X-RAYS

- Require 3 weeks to 2 months postinjury to reveal HO. Useful to confirm maturity of HO.

Heterotopic Ossification Prophylaxis

- ROM exercises
- Control of spasticity
- NSAIDs
- Radiation used perioperatively to inhibit HO in total hip replacement patients; concerns about increased risk of neoplasia limit its use in younger patient populations
  - Radiation in TBI patients for HO prophylaxis would require essentially irradiation of the whole body (as HO can develop practically at any joint), which is not practical.

Treatment

- Bisphosphonates and NSAIDs (particularly indomethacin) have been used on patients to arrest early HO and to prevent postop recurrence, but their efficacy has not been clearly proven in the TBI population.
- ROM exercises are used for prophylaxis and treatment of developing HO to prevent ankylosis.
- Surgical resection of HO indicated only if function is the goal (e.g., hygiene, activities of daily living [ADLs], transfers).
  - Surgical resection usually postponed 12 to 18 months to allow maturation of HO
  - On radiograph, mature HO appears as a well-demarcated radiodense mass with highest radiodensity in the periphery "eggshell calcification" (Meyers et al., 2019).

HYPERTENSION

- Frequently observed post-TBI: Estimated incidence 11% to 25%
- Posttraumatic HTN usually resolves spontaneously. Long-term use of antihypertensive agents is rarely necessary.
- Post-TBI HTN related to sympathetic hyperactivity usually seen in severe TBI—demonstrated by plasma and urine catecholamine levels
- Cases of HTN have been reported secondary to hydrocephalus several years after TBI.

- If medication is needed, propranolol is recommended because it:
  - may reduce the action of catecholamines
  - reduces the cardiac index
  - decreases myocardial oxygen demand
  - decreases heart rate
  - improves pulmonary ventilation-perfusion inequality

VENOUS THROMBOEMBOLIC DISEASE

- Venous thromboembolism (VTE), including DVT and pulmonary embolus (PE), is among the most significant complications of TBI, as it is related to increased mortality in the rehabilitation setting.
- Incidence of DVT in TBI rehabilitation admissions is approximately 10% to 18% (Cifu et al., 1996).
- VTE is often asymptomatic in the TBI population, with sudden death from PE being the first clinical sign in 70% to 80%.
- DVTs occur most commonly in the lower limbs and are traditionally associated with immobility, paresis, fracture, soft tissue injuries, and age >40.
- Remember Virchow's triad: Venous stasis, vessel-wall damage, and hypercoagulable state.

Diagnostic Studies for Deep Vein Thrombosis (Table 2–12)

- Doppler ultrasonography
- Impedance plethysmography (IPG)
- ^{125}I-fibrinogen scanning
- Contrast venography: Gold standard (Carlile et al., 2010)

TABLE 2–12 Diagnostic Tests for DVTs

DIAGNOSTIC TEST	PROS	CONS
Doppler ultrasonography	• 95% sensitivity and 99% specificity for symptomatic proximal thrombi	• Limited ability to detect calf thrombi
IPG	• 90%–93% sensitivity and 94% specificity for proximal thrombi	• Limited ability to detect calf thrombi
^{125}I-fibrinogen scanning	• 60%–80% sensitive in proximal thrombi	• Invasive • Involves injection of radioactive agent
Contrast venography	• Remains the gold standard for diagnosis of clinically suspected DVT	• Invasive • Contrast-induced thrombosis • May be contraindicated if contrast allergy exists

DVT, deep vein thrombosis; IPG, impedance plethysmography.

Deep Vein Thrombosis Prophylaxis in Traumatic Brain Injury

- Chemoprophylaxis:
  - Adequate anticoagulation generally achieved with low-dose unfractionated heparin (5,000 U q 8–12 hours) or low-molecular-weight heparin (LMWH)
- If there is a contraindication to anticoagulation:
  - Intermittent pneumatic compression—provides effective DVT prophylaxis in patients at risk of bleeding complications
  - Inferior vena cava (IVC) filter (Carlile et al., 2010)

Treatment of Venous Thromboembolism

- Therapeutic anticoagulation is first initiated with intravenous (IV) heparin or dose-adjusted subcutaneous (SQ) LMWH, followed by oral anticoagulation (warfarin or direct oral anticoagulants [DOACs]; Julia & James, 2017).

- Anticoagulation continues for 3 to 6 months.
- IVC filter is placed when anticoagulation is contraindicated.

🕮 SYNDROME OF TREPHINED, "SUNKEN FLAP SYNDROME"

(Sveikata et al., 2022)

- Rare complication following craniectomy characterized by delayed sensorimotor or cognitive worsening associated with headache, dizziness, fatigue, and pain at craniectomy site
- Sinking skin flap is theorized to disrupt cerebral blood and CSF flow.
- Neurologic dysfunction improves with cranioplasty.

🕮 CAROTID-CAVERNOUS FISTULA

- Direct connection frequently caused by trauma between internal carotid artery and cavernous sinus
- Symptoms include diplopia, tearing, red eye, headache, proptosis, and subjective ocular bruit.
- Treatment: Endovascular embolization of fistula

URINARY DYSFUNCTION

- Uninhibited neurogenic bladder with uninhibited detrusor reflex (contraction)
- TBI patients are frequently incontinent, usually presenting with an uninhibited type neurogenic bladder, in which the bladder volume is reduced but empties completely with variable postvoiding intravesicular residual (PVR) volumes (Dorsher & McIntosh, 2012).
- For this type of dysfunction, a time-void program is usually helpful, in which the patient is offered the urinal or commode at a regularly scheduled interval.
- Anticholinergic meds (decreases detrusor tone → increases bladder capacity) may also be used.
- Note: For a more detailed description of bladder function, types of neurogenic bladder, and treatments, see Chapter 7, Spinal Cord Injury.

SPASTICITY

- Disorders of abnormal motor tone (e.g., spasticity, rigidity) are common after TBI.
- Acute TBI and the resulting increase in muscle tone cause a state of hypermetabolism, which increases energy requirements from 100% to 140% of what is expected.
- Please refer to the Spasticity section in the Associated Topics chapter for a full discussion on definition, clinical assessment/grading, and treatment options for spasticity.

NUTRITION

- TBI patients generally have higher caloric and protein requirements due to hypermetabolism, increased energy expenditure, and increased protein loss.
- Measured resting energy expenditure (mREE) after TBI varies, ranging from 125% in coma to 250% with PAID syndrome (Kurtz & Rocha, 2020).
- 🕮 Initiate nutrition within 24 hours (>50% mREE) and advance quickly to >80% over 1 week with a protein intake goal of up to 1.5 g protein/kg over 2 weeks (Kurtz & Rocha, 2020; Tavarez et al., 2021).
- For many TBI patients, swallowing is inhibited by both cognitive and oral motor deficits and may require alternate feeding routes (enteral or parenteral).
- Nutritional status monitoring via routine blood work and frequent weight checks is necessary, and the potential for oral feeding should routinely be reassessed. Thorough dysphagia management should include a specially trained therapist, clinical dietitian/nutritionist, and an eventual oral motor facilitation treatment program. Useful diagnostic tools include video fluoroscopy, performed by an experienced therapist and radiologist, and occasionally fiberoptic endoscopy (Spiegel et al., 1998).

- Failure to wean patients off of alternate feeding routes has been associated with persistently poor intraoral manipulation and with cognitive levels below V on the Ranchos Los Amigos Scale.

🕮 Enteral Feeding

- Preferred when oral feeding is compromised because it directly uses the GI tract (distal to the site of tube placement), provides the most physiologic approach in nutritional administration and absorption, is low in cost, and has a lower risk of metabolic complications
- The primary risk for tube feedings is aspiration, and this risk is increased with gastroesophageal reflux disease (GERD) or with more proximal tube placement. Risks with distal tube placement include decreased absorptive capacity and tolerance of the remaining gut.
- Enteral feeding products include pureed foods, liquid nutritional supplements, elemental nutritional supplements, or a combination of products.
- Enteral routes include nasogastric, nasoenteric, esophagogastric, percutaneous placement (gastrostomy, jejunostomy), and more surgically invasive tubes (Janeway gastrostomy, esophagogastrostomy).
- Currently, there are no guidelines dictating how soon a feeding tube should be placed or the optimal location of tube placement (gastrostomy vs. jejunostomy), but several factors should be taken into consideration.
  - Direct gastrostomy or jejunostomy has a decreased risk of aspiration and GERD-related problems; they are preferred when there is a potentially prolonged length of time of nonoral nutrition (Grahm et al., 1989).
  - These direct routes should be in place for at least 30 days to decrease the complications associated with removal.
  - Percutaneous tube placement has the added advantages of lower surgical risks and the ability to start tube feeds within 24 hours of placement, whereas more surgically placed tubes have mechanical parts that can more easily be inserted (during mealtimes only) and removed (especially when in therapy; Kirby et al., 1991).
  - Enteral routes that allow for bolus feeding are advantageous because they more closely approximate natural feeding, making daily routines and therapies more manageable, especially in patients likely to go home.
- Associated problems:
  - In patients with GERD, recurrent pneumonia, or possible aspiration, distal tube placement is preferred.
  - Patients suspicious for aspiration or aspiration pneumonia should have a gastric source of aspirate confirmed to rule out the aspiration of oral secretions.
  - GERD has a high prevalence in TBI patients and can also lead to aspiration and esophagitis.
  - Head elevation may reduce the risk of aspiration, and antacids may improve esophagitis.
  - A high level of gastric residue was the most common feeding intolerance found in TBI patients, and the delivery of erythromycin by NGT may control GI disorders.
  - Although metoclopramide (Reglan®), a dopamine antagonist, increases gastroesophageal sphincter tone and can aid in GERD, it should be avoided due to its ability to cause sedation, potential to impair recovery, and its extrapyramidal side effects.

🕮 Parenteral Feeding

- Intravenously delivered nutrition, usually through a central venous line, or, in limited circumstances, a peripheral line
- Parenteral feeding can be either supplemental or primary (total parenteral nutrition [TPN]).
  - Parenteral supplementation is utilized when there is a temporary interruption of GI function or in any condition with an increased metabolic demand.
  - TPN is preferred when a segment of GI tract is nonfunctional or must be free of food for a prolonged amount of time.

- Because parenteral feeding products bypass essential GI metabolism, they are made of a constitution of nutrients that must be in an elemental form. Optimal proportions of elements within a parenteral solution vary widely and should frequently be reassessed.
- Risks of parenteral feeding include central/peripheral line complications (infection, clot formation, edema). Central lines have an added risk of pneumothorax during catheter insertion. Electrolyte and metabolic abnormalities are common with parenteral feeding and should be closely monitored.

NEUROENDOCRINE DISORDERS AFTER TRAUMATIC BRAIN INJURY

Hypothalamic Pituitary Dysfunction

- Two-thirds of severe TBI mortalities with structural abnormalities occur in the hypothalamic pituitary region.
- Originally thought to be a rare condition, more recent evidence suggests that a much higher percentage of TBI patients have anterior pituitary dysfunction. Some studies show rates as high as 50%, greater in patients with severe TBI relative to mild-moderate injury.
- Growth hormone deficiency (GHD) is the most common abnormality, followed by hypogonadism, hypothyroidism, hypocortisolism, and diabetes insipidus (Schneider et al., 2007).
- Growth hormone (GH) is secreted in a pulsatile fashion and insulin growth factor 1 (IGF-1) is only correlated with GH levels.
- Ghigo et al. (2005) have suggested an algorithm for the evaluation and treatment of people who have sustained a TBI.
- Recommend that all patients undergo endocrine function evaluation at 3 months and at 1 year postinjury regardless of injury severity
- Recommended screening: AM cortisol, IGF-1, follicle-stimulating hormone (FSH), luteinizing hormone (LH), testosterone, estradiol, prolactin, and urinary free cortisol
- Based on the screening studies, more involved provocative testing may be ordered and possible hormone replacement.
- Please refer to Ghigo et al. (2005) for more details.

PATHOPHYSIOLOGY

- Direct and indirect trauma to the brain
- Drugs
- Circulating cytokines
- Secondary insults
- Vascular injury

Hyponatremia

- Hyponatremia in TBI is generally present in a hypotonic setting with either normal extracellular volume (isovolemia = syndrome of inappropriate antidiuretic hormone secretion [SIADH]) or reduced extracellular volume (hypovolemia = cerebral salt wasting [CSW]).
- It is important to understand the different causes of hyponatremia, as treatments for each condition are markedly different. This is also particularly important as hyponatremia may also cause cognitive dysfunction.

Syndrome of Inappropriate Antidiuretic Hormone Secretion (Table 2–13)

- Water retention resulting from excessive antidiuretic hormone (ADH) secretion from the neurohypophysis that is secondary to multiple causes including head trauma
- In SIADH, ADH excess is considered inappropriate because it occurs in the presence of plasma hypo-osmolality and despite normal or increased plasma volume (i.e., euvolemic hyponatremia).

- In SIADH, Na^+ excretion in the urine is maintained by hypervolemia, suppression of the renin–angiotensin–aldosterone system, and ↑ in the plasma concentration of atrial natriuretic peptide (usually >20 mmol/L).

COMMON CAUSES OF SYNDROME OF INAPPROPRIATE ANTIDIURETIC HORMONE SECRETION

- **CNS diseases:**
  - Thrombotic or hemorrhagic events
  - Infection
  - Meningitis
  - Encephalitis
  - Brain abscess
  - CNS neoplasm
- **Head trauma**
- **Lung disease:**
  - Pneumonia
  - Lung abscess
  - Positive pressure ventilation
- **Malignancy:**
  - Cancer (CA) of the lung (especially small cell CA)
  - GI malignancy (e.g., pancreatic CA)
  - Prostate CA
  - Thymoma
  - Lymphoma
- **Drugs:**
  - 🕮 Carbamazepine
  - Vincristine
  - Clofibrate
  - Chlorpropamide
  - Phenothiazines
  - Amitriptyline
  - Morphine
  - Nicotine
  - SSRIs

SIGNS AND SYMPTOMS OF SYNDROME OF INAPPROPRIATE ANTIDIURETIC HORMONE SECRETION

- In mild SIADH (with Na^+ 130–135), or in gradually developing SIADH, symptoms may be absent or limited to anorexia and nausea/vomiting.
- In severe SIADH (with significant hyponatremia) or in acute-onset SIADH, there might be an increase in body weight and symptoms of cerebral edema—restlessness, irritability, confusion, convulsions, and coma.
- Edema (peripheral/soft tissue) is almost always absent.

🕮 TREATMENT

- Fluid restriction to approximately 1.0 L/day (800 mL–1.2 L/day; either alone or with a loop diuretic)
- Careful daily monitoring of weight changes and serum Na^+ until sodium level >135 mmol/L
- Hypertonic saline (e.g., 3% NaCl solution)—200 to 300 mL should be infused IV over 3 to 4 hours in patients with severe symptoms as confusion, convulsions, or coma.
- 🕮 It is important *not* to raise Na^+ concentration too rapidly to avoid development of serious neurologic damage, pontine myelinolysis, or congestive heart failure (CHF). Sodium may be corrected no more than 10 mEq/L over 24 hours until sodium levels reach 125 mEq/L. NaCl repletion with salt tablets.
- Tolvaptan is a selective vasopressin V2-receptor antagonist approved for treating hyponatremia in SIADH (Bondanelli et al., 2023).
- Chronic SIADH may be treated with demeclocycline, which normalizes serum Na^+ by inhibiting ADH action in the kidney; lithium carbonate acts similarly but is rarely used because it is more toxic.

Cerebral Salt Wasting Syndrome (Table 2–13)

- CSW is another common cause of hyponatremia in TBI. It may be a more common cause of hyponatremia in TBI patients than SIADH.

- CSW is thought to occur because of direct neural effect on renal tubular function.
- In CSW, hyponatremia is not dilutional (as in SIADH)—CSW patients are, in fact, volume depleted.
- *Hallmark* of CSW:
  - Decreased blood volume (↓ extracellular volume = hypovolemia) secondary to sodium loss (in urine), triggering increased ADH secretion that is appropriate rather than inappropriate (differentiating this condition from SIADH).
  - Signs of dehydration are present.

TREATMENT OF CEREBRAL SALT WASTING

- Hydration/fluid replacement + electrolyte (Na^+) correction
- It is important to differentiate CSW from SIADH and to recognize that there is water depletion in this condition, as treating it with fluid restriction (appropriate treatment for SIADH) may further reduce the extracellular fluid with disastrous results to the patient.

Psychogenic Polydipsia

- Behavioral disorder seen rarely in people with TBI
- Polydipsia with hyponatremia
- Behavioral, dopaminergic, and cholinergic systems as well as hippocampal pathology
- Treatment: Behavioral modification, fluid restriction, and clozapine

Diabetes Insipidus (Table 2–13)

- Diabetes insipidus (DI) represents a deficiency of ADH (vasopressin).
- In contrast to SIADH or CSW, hypernatremia can result due to excessive volume depletion.
- It may occur in severe head injuries and is often associated with fractures of the skull.
  - 🕮 A fracture in or near the sella turcica may tear the stalk of the pituitary gland, resulting in DI (due to disruption of ADH secretion from posterior pituitary), in addition to other clinical syndromes, depending on the extent of the lesion.
- Spontaneous remissions of traumatic DI may occur even after 6 months, presumably because of regeneration of disrupted axons within the pituitary stalk.

TABLE 2–13 Comparison of Diagnostic Labs and Treatment of SIADH, DI, and CSW

	SIADH	DI	CSW SYNDROME
Serum ADH (rarely done as routine lab work)	↑ ("inappropriately" elevated)	↓	↑ ("appropriately" elevated)
Serum Na^+	↓	↑	↓
Serum osmolality	↓	↑	↓
Extracellular volume	Normal (isovolemic)	Normal (isovolemic)	Reduced (hypovolemic)
Urine osmolality and SG	↑ (concentrated urine with osmolality usually >300 mmol/kg)	↓	Normal
Treatment	Fluid restriction, give Na^+, Vaprisol, demeclocycline, tolvaptan	DDAVP	Give volume, give Na^+, fludrocortisone

ADH, antidiuretic hormone; CSW, cerebral salt wasting; DI, diabetes insipidus; SG, specific gravity; SIADH, syndrome of inappropriate antidiuretic hormone secretion.

CLINICAL MANIFESTATIONS

- Polyuria, excessive thirst, and polydipsia
- Urinary concentration (osmolality <290 mmol/kg, specific gravity <1.010) is below that of the serum in severe cases but may be higher than that of serum (290–600 mmol/kg) in mild DI.
- Normal function of the thirst center ensures that polydipsia closely matches polyuria, so dehydration is seldom detectable except by a mild elevation of serum Na^+.
- However, when replenishment of excreted water is inadequate, dehydration may become severe, causing weakness, fever, psychic disturbances, prostration, and death.
- These features are associated with a rising serum osmolality and serum Na^+ concentration, the latter sometimes >175 mmol/L.

TREATMENT

- DDAVP (desmopressin acetate)—ADH analog with prolonged antidiuretic effect and no significant pressor activity. May be given intranasally or IM.
- Chlorpropamide potentiates the effects of ADH on the renal tubules—used in partial ADH deficiency.

COGNITIVE DYSFUNCTION

- Numerous cognitive issues arise as a result of TBI. They include problems with attention, executive control, encoding, and recall of new memory, as well as self-monitoring.
- 🕮 There is a link between TBI severity and rate of cognitive decline. Alzheimer's dementia (AD) is strongly associated with moderate and severe TBI but not mild TBI without loss of consciousness (LOC; Bazarian et al., 2009):
  - There is a tenfold increased risk of AD with both apolipoprotein-epsilon 4 (APOE4) genotype and a history of traumatic head injury (Mayeux et al., 1995).
- *Cognitive rehabilitation*: Comprehensive, holistic approach that attempts to address multiple cognitive deficits and incorporates psychological interventions for emotional, motivational, and interpersonal aspects of the patient's functioning
- High return to work rates were associated with higher preinjury educational level of functioning, premorbid functional status, and work opportunities postinjury.
- Intensive, holistic cognitive remediation programs showed better community reintegration compared with those with a "standard" rehabilitation program.
- 🕮 Cognitive remediation includes visuospatial rehabilitation, executive control, self-monitoring, pragmatic interventions, memory retraining, and strategies to improve attention.
- Specific interventions for attention, memory, and executive functioning demonstrated benefits, although subject sizes were limited.
- Compensatory techniques for pressure management or memory deficits proved effective, although subject sizes were limited.
- A 2005 literature review by Cicerone et al. (2005) reported that over 28% of studies noted the efficacy of cognitive remediation over control.

Pharmacologic Interventions for Specific Cognitive Deficits

- Literature is still limited. Most recent reviews (Chew & Zafonte, 2009; Gordon et al., 2006; Warden et al., 2006) have given some general guidance. It is critical to identify a clear target for intervention.

Arousal and Attention

- Literature has shown the efficacy of methylphenidate, amantadine, bromocriptine, and levodopa in increasing arousal and attention (Giacino et al., 2013).

- Methylphenidate has also shown to be of benefit for processing speed.
- Acetylcholinesterase inhibitors have also been suggested as a potentially effective agent for these problems. Evidence-based reviews suggest these agents as potential treatment options.

Memory

- There is even less evidence-based literature for the use of medications in the management of memory dysfunction.
- Some evidence suggests that the cholinesterase inhibitors may be beneficial, and greater evidence exists for donepezil.
- Methylphenidate and cytidine diphosphocholine may also be considered as treatment options.
- Numerous other agents have been trialed, often working on the cholinergic or catecholaminergic pathways. However, limited evidence for efficacy is currently available.

Aphasia

- Aphasia is a problem often seen after TBI, with Sarno reporting a rate of between 2% and 32% in the civilian population, while a more recent study of veterans reported a rate of 1.5% (Lindsey et al., 2023; Sarno, 1984).
- Donepezil has been used in multiple populations with improvements in language function, and recently in a case report of a person with TBI, so its use should be considered (Swenson et al., 2022).

Guidelines for Pharmacologic Intervention

- Start low and go slow.
- Provide an adequate therapeutic trial.
- Perform continuous reassessment.
- Monitor drug–drug interactions.
- Consider drug augmentation.
- Change strategy if symptoms intensify.

MILD TRAUMATIC BRAIN INJURY (CONCUSSION) AND POSTCONCUSSIVE SYNDROME

MILD TRAUMATIC BRAIN INJURY (CONCUSSION)

- Mild TBI constitutes 80% to 90% of TBI cases in the United States (approximately 2.3 million cases).
- Multiple terms, definitions, and diagnostic criteria are available for mild TBI.
- The American Congress of Rehabilitation Medicine (Giacino et al., 1995) defined mild TBI as a traumatically induced physiologic disruption of brain function with at least one of four manifestations:
  - Any LOC
  - Any loss of memory of events immediately before or after the injury
  - Any alteration in mental status at the time of the accident
  - Focal neurologic deficits that may or may not be transient
- The injury **does not exceed** the following severity criteria:
  - LOC of 30 minutes
  - PTA of 24 hours
  - Initial GCS of 13 to 15

- Usually, mild TBI has no findings of structural injury on routine neuroimaging (CT/MRI).
- Signs and symptoms after mild TBI include:
  - Headache (most common)
  - Dizziness
  - Tinnitus
  - Impaired balance
  - Hearing loss
  - Blurred vision
  - Altered taste and smell
  - Sleep disturbances/insomnia
  - Fatigue
  - Sensory impairments
  - Attention and concentration deficits
  - Slowed mental processing (slowed reaction and information processing time)
  - Memory impairment (mostly recent memory)
  - Lability
  - Irritability
  - Depression
  - Anxiety
- Most mild TBI patients have a good recovery, with symptoms clearing within the first few weeks or months postinjury (usually within 1–3 months).
- Pharmacologic intervention may be used, including antidepressants and psychostimulants.

Second Impact Syndrome

- Results from a person (usually an athlete) sustaining a second brain injury (that may be minor in severity) before symptoms of a prior concussion have cleared. Immediately following the second head injury, patients become dazed, and within 15 seconds to several minutes can rapidly decompensate—collapse, pupil dilation, loss of eye tracking, respiratory failure, and semicomatose state.
- Current research suggests an impairment in the brain's vascular autoregulation, leading to engorgement and increased ICP that result in herniation (either of the medial temporal lobe through the tentorium or the cerebellar tonsils through the foramen magnum).
- Incidence of second impact syndrome is unknown and likely underreported, but studies suggest morbidity and mortality rates close to 100% and 50%, respectively.
- More common in adolescent-aged athletes

🕮 POSTCONCUSSION SYNDROME

- In the *International Classification of Diseases*, Tenth Edition *(ICD-10;* 1992*)* criteria, postconcussion syndrome (PCS) includes:
  - A history of head trauma with LOC preceding symptom onset by a maximum of 4 weeks
  - There must be symptoms in at least three or more of the following categories:
    - Headaches
    - Noise intolerance
    - Dizziness
    - Malaise
    - Fatigue
    - Light intolerance
    - Irritability
    - Anxiety

    - Depression
    - Emotional lability
    - Subjective concentration, memory, or intellectual difficulties without neuropsychological evidence of impairment
    - Reduced alcohol tolerance
    - Preoccupation with the previously mentioned symptoms
  - Prolonged symptom duration, as well as defining subjective symptoms and objective findings, remains controversial.
- The *Diagnostic and Statistical Manual of Mental Disorders*, Fifth Edition (*DSM-5*) refers to the major or mild neurocognitive disorder due to TBI:
  - The *ICD* refers to postconcussive state as a syndrome, but the *DSM-5* does not.
  - Neurocognitive symptoms associated with mild TBI are noted to resolve within days to weeks after the injury, with complete resolution by 3 months according to the American Psychiatric Association (2013).
- PCS is associated with social and vocational difficulties that appear to be out of proportion to the severity of the neurologic insult.
  - Persistent PCS has been used to describe symptoms lasting over 3 to 6 months.

CONCUSSION CATEGORIZATION

- Concussions are no longer categorized on severity scales. Classification systems were cast aside in 2013 and are now based on clinical judgment alone.
- Severity is now gauged using a symptom severity scale, such as *Sport Concussion Assessment Tool*, Sixth Edition (SCAT6) or Acute Concussion Evaluation (ACE).
- SCAT6 is a standardized tool developed by the Concussion in Sport Group used to evaluate concussive injury in athletes 13 years of age and older.
- The SCAT6 includes an on-field and off-field evaluation.
- The on-the-field evaluation includes observable signs, GCS, cervical spine examination, coordination and ocular motor examination, and a basic memory examination called the Maddocks questions.
- There are also red flag items that prompt immediate removal from play, including neck pain or tenderness, seizures, double vision, LOC, weakness or tingling in more than one extremity, deteriorating level of consciousness, vomiting, severe or increasing headache, increased restlessness, agitative or combative GCS <15, and visual deformity of the skull.
- Off-the-field evaluation includes athlete background, symptom evaluation, cognitive screen, and a coordination and balance evaluation.
- The ACE is an initial assessment tool for concussion that the public can use.
- The clinician generates a total score of severity and counts the domains in which the person has symptoms. A consensus statement following the International Sports Concussion Conference in Zurich developed a symptom checklist composed of four different domains.
- The four domains are:
  1. Somatic (headache, dizziness, visual disturbances, nausea, and the like)
  2. Cognitive (confusion, LOC, inability to concentrate, and memory problems)
  3. Affective (emotional lability, anxiety, sadness, and irritability)
  4. Sleep changes (trouble falling asleep, or sleeping more or less than usual)
- The combination of symptoms reported by the patient and a neurologic exam, in which the doctor evaluates the patient's vital signs, visual performance, balance, memory, and cognitive functioning, guides how the concussion is managed.
- Treatment for concussions includes physical and cognitive rest for 48 hours, followed by a gradual return to activities, learning, or work, in addition to a graded aerobic exercise regimen under clinical supervision.

GUIDELINES FOR RETURN TO SPORTS AFTER CONCUSSION (TABLE 2–14)

- Return-to-sports (RTS) criteria have been similarly controversial. The Colorado Medical Society and Cantu guidelines are widely used.
- In 2022, the Sixth International Conference on Concussion in Sport released its most recent Consensus Statement on Concussion in Sport.
  - Assessment of concussion references the SCAT6 and the Child-SCAT6 (Sport Concussion Assessment Tool for children 8 to 12 years old).
  - It explicitly recommends no RTS on the day of a concussive injury regardless of the severity.
  - Concussion management and RTS guidelines begin with physical and cognitive rest until the acute symptoms resolve (usually 24–48 hours), followed by a stepwise graded program of exertion prior to medical clearance and RTS.
  - In the Graduated Return to Sport Protocol, the athlete can only proceed to the next level if asymptomatic at the current level. Generally, each step should take at least 24 hours, thereby resulting in at least 1 week to proceed through the full rehabilitation protocol. If any postconcussion symptoms occur, the athlete is to drop back to the previous asymptomatic level and try to progress again after another 24-hour period of rest has passed.

TABLE 2–14 Return to Sports Protocol From the Sixth International Conference on Concussion in Sports, 2022

REHABILITATION STAGE	FUNCTIONAL EXERCISE AT EACH STAGE OF REHABILITATION	OBJECTIVE OF EACH STAGE
1. Symptom-limited activity	Daily activities that do not provoke symptoms	Gradual reintroduction of school/work activities
2. Aerobic exercise (2A-light and 2B-moderate) 2A—Light (up to approximately 55% maxHR) then 2B—Moderate (up to approximately 70% maxHR)	Stationary cycling or walking at slow to medium pace • Keeping intensity <55% maximum permitted HR for 2A, <70% for 2B No resistance training	Increase HR
3. Sport-specific exercise	Skating drills in ice hockey, running drills in soccer; no head impact activities	Add movement, change of direction
4. Noncontact training drills	Progression to more complex training drills (e.g., passing drills in football and ice hockey) May start progressive resistance training	Resume usual intensity of exercise, coordination, and cognitive load
5. Full-contact practice	Following medical clearance, participate in normal training activities	Restore confidence and assess functional skills by coaching staff
6. Return to sport	Normal game play	

Note: A symptom-free period of at least 24 hours is required prior to progressing the athlete to the next rehabilitation stage. Athletes should not be progressed if they are still symptomatic and should regress back to the rehabilitation stage where they are asymptomatic. Student athletes should complete full return-to-learn protocol before unrestricted return-to-sport.
HR, heart rate.
Source: From Patricios JS, Schneider KJ, Dvorak J, et al. Consensus statement on concussion in sport: the 6th International Conference on Concussion in Sport-Amsterdam, October 2022. *Br J Sports Med*. 2023;57(11):695–711. doi:10.1136/bjsports-2023-106898.

CHRONIC TRAUMATIC ENCEPHALOPATHY (Mckee et al., 2016)

- A neurodegenerative disease caused by repetitive brain trauma leading to diffuse hyperphosphorylated tau accumulation.
- Chronic traumatic encephalopathy (CTE) is documented in athletes involved in contact sports (football, ice hockey, boxers, and wrestling) and military personnel exposed to blast injuries.
- Clinical presentations are variable favoring between behavioral/mood changes and cognitive impairment.
  - Behavior/mood group: Symptoms present at younger age, demonstrate impulsivity, physical and verbal violence, and depression
  - Cognition group: Demonstrate episodic memory impairment, more likely to progress to dementia, older at age of death relative to behavior/mood group

CENTRAL NERVOUS SYSTEM CONDITIONS SECONDARY TO HIV

- 30% to 50% of HIV-positive patients have neurologic complications secondary to the disease.

TOXOPLASMOSIS

- Caused by infection with the protozoan *Toxoplasma gondii*
- *Toxoplasma encephalitis* presents in patients with CD4 T-lymphocyte counts <200.
- Most common symptoms include headache, confusion, fever, and lethargy. Presentation is usually subacute, from a few days to a few months.
- Imaging reveals enhancing lesions surrounded by edema.
- Treatment: Combination of pyrimethamine and sulfadiazine

CRYPTOCOCCAL MENINGITIS

- This is the most common opportunistic meningitis in HIV.
- Infects HIV patients with CD4 counts <200
- Patients show acute signs of fever, headache, confusion, and meningeal irritation signs.
- Treatment: Amphotericin B

CENTRAL NERVOUS SYSTEM LYMPHOMA

- Most common HIV-associated brain malignancy. Associated with Epstein–Barr virus (EBV)
- CD4 counts are very low at the time of diagnosis.
- Common symptoms are confusion, lethargy, and memory impairments.
- Oral or IV steroids can be considered for those with altered mental status, focal symptoms, or impending herniation for temporary improvement.
- Treatment: Whole brain irradiation; steroids are added to reduce edema and mass effect.

AIDS DEMENTIA

- Occurs in 15% of patients with AIDS and can be the first manifestation of the disease in 3% to 10% of patients
- Clinical characteristics can be broken down into three subdivisions: Cognition, behavioral, and motor.
  - Cognition: Forgetfulness, mental slowing, and decreased concentration
  - Behavioral: Apathy and social withdrawal
  - Motor: Gait instability and poor coordination
- Treatment: Antiretroviral therapy (ART; prevents viral replication and increasing viral load)

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RHEUMATOLOGY

3

Thomas R. Nucatola, MD • Eric D. Freeman, DO • Leslie Bagay, MD •
David P. Brown, DO • Seema Malkana, DO • Jaclyn Joki, MD • Faraz Jamal, DO

RHEUMATOID ARTHRITIS

INTRODUCTION

Rheumatoid arthritis (RA) is a systemic autoimmune inflammatory disorder of unknown etiology affecting multiple organ systems. In the joints of the musculoskeletal system, it primarily affects the synovial lining of diarthrodial joints. This chronic, symmetric erosive synovitis develops in the joints and leads to articular destruction. These erosions are pathognomonic of RA (Kelly et al., 1997; Klippel, 1997).

Joint Destruction in Rheumatoid Arthritis

- Initially starts with injury to synovial microvasculature
- Fibroblast-like synoviocytes are activated via class III human leukocyte antigens (HLAs; cellular process), which leads to synovial cell proliferation.
- Leads to congestion, edema, and fibrin exudation
- T-lymphocytes infiltrate
- The synovium then hypertrophies (cartilage is destroyed).
- Pannus formation occurs.

Composition of Diarthrodial Joints
- Type II hyaline cartilage
- Subchondral bone
- Synovial membrane
- Synovial fluid
- Joint capsule

🕮 Pannus Formation

- Pannus formation is the most important destructive element in RA.
- Pannus is a membrane of granulation tissue that covers the articular cartilage at joint margins. Pannus is a synovial membrane-derived tissue that overgrows cartilage.
- Fibroblast-like and macrophage-like cells invade and destroy the periarticular bone and cartilage at joint margins.
- Osteoclasts invade bone, leading to marginal erosions at the pannus–bone interface.
- Vascular granulation tissue is composed of:
  - Proliferating fibroblasts
  - Numerous small blood vessels
  - Various inflammatory cells, but mainly CD4+ T-lymphocytes. Note that polymorphonuclear cells (PMNs) predominate in synovial fluid.
  - Occasionally, collagen fibers are seen within phagolysosomes of cells at the leading edge of pannus.

- Pannus is vascular (angiogenesis).
  - CD4 and T-cells predominate in pannus (but PMNs are in synovial fluid).
  - Cytokines mediate inflammation (interleukin [IL-1], tumor necrosis factor alpha [TNF-alpha], IL-6, IL-17, granulocyte-macrophage colony-stimulating factor [GM-CSF], and tumor growth factor beta [TGF-beta]).
- Joint ankylosis may occur in later stages.

EPIDEMIOLOGY

- Female-to-male ratio is 2:1.
- Prevalence: Approximately 1% of the population worldwide and about 1.3 million U.S. adults (Helmick et al., 2008)
- Genetic:
  - Major histocompatibility complex (MHC) on chromosome 6
  - Class II MHC allele HLA-DR4 (HLA-DR4 haplotype)
- Age range: 20 to 60 years of age; prevalence rises with age; peak incidence between fourth and fifth decades

ETIOLOGY

- The precise cause of RA is unknown.
- Contributing factors include:
  - Environmental factors (cigarette smoking)
  - Socioeconomic factors: Low socioeconomic status is associated with increased prevalence of RA, increased time to diagnosis, and poorer outcomes.
  - Hormonal

🕮 CLINICAL DIAGNOSIS OF RHEUMATOID ARTHRITIS

- Prior American College of Rheumatology (ACR) guidelines for the diagnosis of RA were based on a 1988 American Rheumatologic Association paper by Arnett et al. (1988). In 2010, the ACR and European League Against Rheumatism (EULAR) created new classification criteria for RA (Aletaha et al., 2010) to capture earlier presentation of symptoms.

2010 AMERICAN COLLEGE OF RHEUMATOLOGY/EUROPEAN LEAGUE AGAINST RHEUMATISM CLASSIFICATION CRITERIA FOR RHEUMATOID ARTHRITIS

- To apply this criteria, the patient must have at least one joint with definite clinical synovitis (swelling) that is not best explained by another disease process.
- **A score of ≥6/10 is needed for classification of a patient as having definite RA.** (See the following table with ACR/EULAR Classification Criteria for RA.)

PATTERNS OF RHEUMATOID ARTHRITIS ONSET

1. 🕮 **Insidious → 50% to 70%**
   - Initial symptoms can be systemic or articular
   - Slow onset from weeks to months
   - Constitutional symptoms: Fatigue, malaise
   - Diffuse musculoskeletal pain may be the first nonspecific complaint with joint involvement later
   - Most commonly symmetric involvement, although asymmetric involvement may be seen early
   - Morning stiffness in the involved joints lasting 1 hour or more
   - Joint swelling, erythema
   - Muscle atrophy around the affected joints
   - Low-grade fever without chills

2. **Acute onset → 10% to 20%**
   - Onset over several days
   - Less symmetric in presentation
   - Severe muscle pain
3. **Intermediate onset → 20% to 30%**
   - Onset over several days to weeks
   - Systemic complaints more noticeable

2010 ACR/EULAR Classification Criteria for RA

A. JOINT INVOLVEMENT	SCORE
1 large joint	0
2–10 large joints	1
1–3 small joints	2
4–10 small joints	3
>10 joints (with at least one small joint)	5
B. SEROLOGY (AT LEAST ONE TEST RESULT IS NEEDED FOR CLASSIFICATION)	**SCORE**
Negative RF *and* negative ACPA	0
Low-positive RF *or* low-positive ACPA	2
High-positive RF *or* high-positive ACPA	3
C. ACUTE PHASE REACTANTS (AT LEAST ONE TEST RESULT IS NEEDED FOR CLASSIFICATION)	**SCORE**
Normal CRP *and* normal ESR	0
Abnormal CRP or abnormal ESR	1
D. DURATION OF SYMPTOMS	**SCORE**
<6 weeks	0
≥6 weeks	1

ACPA, antibodies to cyclic citrullinated peptides; ACR, American College of Rheumatology; CRP, C-reactive protein; ESR, erythrocyte sedimentation rate; EULAR, European League Against Rheumatism; RA, rheumatoid arthritis; RF, rheumatoid factor.
Source: Adapted from Aletaha D, Neogi T, Silman AJ, et al. Rheumatoid arthritis classification criteria: an American College of Rheumatology/European League Against Rheumatism collaborative initiative. *Arthritis Rheum.* 2010;62(9):2569–2581. doi:10.1002/art.27584.

🕮 Morning Stiffness of Major Arthritides: Duration and Location

ARTHRITIS TYPE	JOINT(S) INVOLVED	MORNING STIFFNESS DURATION
Rheumatoid arthritis	MCP, PIP, MTP joints	>1 hour
Osteoarthritis	Distal interphalangeal joints, knees, hips	<30 minutes
Ankylosing spondylitis	Lumbosacral spine	Approximately 3 hours

MCP, metacarpophalangeal; MTP, metatarsophalangeal; PIP, proximal interphalangeal.

LAB TESTING FOR RHEUMATOID ARTHRITIS

- No single test is definitive in diagnosing RA.
- Typical laboratory findings in active disease include:
  - Rheumatoid factor (RF; +; see details later)
  - Antibodies to cyclic citrullinated peptides (ACPA/anti-CCP [anti-cyclic citrullinated peptides]): Specific for RA and correlated with aggressive disease
  - Elevated acute phase reactants: Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP)
  - Complete blood count (CBC): Thrombocytosis, hypochromic microcytic anemia, and eosinophilia
  - Synovial fluid analysis (see box)
- Hypergammaglobulinemia
- Hypocomplementemia

📖 Synovial Fluid Testing in RA

- Low viscosity
- WBC = 1,000–75,000/mm^3
- 70% PMNs
- Transparent—cloudy

PMN, polymorphonuclear; RA, rheumatoid arthritis; WBC, white blood cell.

Rheumatoid Factor in Rheumatoid Arthritis

- 📖 70% to 80% of patients with RA are RF (+).
- The remainder of patients with RA are RF (−). ACR criteria are fulfilled with other positive findings (see the preceding table with the ACR/EULAR Classification Criteria for RA).
- RF (+) is associated with increased severity of disease with increased systemic manifestation.
- Serial titers are of no value.
- RF (+) can be seen in other diseases: Rheumatic (systemic lupus erythematosus [SLE], scleroderma, Sjögren's), viral, parasitic, bacterial, neoplasms, hyperglobulinemic, cryoglobulinemia, and hepatitis C.

Anti-Cyclic Citrullinated Peptide Antibodies (Klippel et al., 2008)

- ACPA are seen in RA, with sensitivity of 80% and specificity of 90% to 95%.
- ACPA are rarely seen in other diseases (psoriatic arthritis, tuberculosis [TB], autoimmune hepatitis).

Increased Erythrocyte Sedimentation Rate and C-Reactive Protein

- ESR and CRP are acute phase reactants and are markers of inflammation.
- However, they are nonspecific inflammatory markers and are not used to diagnose a condition that is nonspecific for RA.

📖 RADIOGRAPHIC FINDINGS IN RHEUMATOID ARTHRITIS (SEE TABLE 3–3)

- Early findings:
  - Soft tissue swelling
  - Joint space widening
- Late findings:
  - Uniform joint space narrowing due to loss of articular cartilage (hips, knees, etc.)
  - Axial migration of the hip (protrusio acetabuli)
  - Malalignment and fusion of joints
- (+) Juxta-articular osteopenia (bone washout)
- Marginal bone erosions (near attachment of joint capsule)
- Predilection of swelling of joints in wrists: Metacarpophalangeals (MCPs), proximal interphalangeals (PIPs), metatarsophalangeals (MTPs) but not distal interphalangeals (DIPs)
- Erosion of the ulnar styloid
- Rupture of radioulnar ligament (piano key sign)
- Erosion of the metatarsal head of the MTP joint
- Disease may be asymmetric at first, then progress to be symmetric.
- Cervical spine involvement may lead to cervical atlantoaxial (A-A) subluxation (>2.5–3 mm; Martel, 1961; Park et al., 1979).

- Ulnar deviation and volar subluxation seen at the MCP joint of the phalanges
- Radial deviation of the radiocarpal joint
- Hallux valgus

JOINT DEFORMITIES IN RHEUMATOID ARTHRITIS

DISEASE PROGRESSION

1. Morning stiffness >1 hour → universal feature of synovial inflammation
2. Structural inflammation → warm swollen tender joints seen superficially
3. Structural damage → cartilage loss and erosion of the periarticular bone

Joints Commonly Involved in RA

- Hands and wrist
- Cervical spine—C1 to C2 → atlantoaxial subluxation
- Feet and ankles
- Hips and knees

RA, rheumatoid arthritis.

UPPER EXTREMITY DEFORMITIES IN RHEUMATOID ARTHRITIS

🕮 Hand and Wrist Deformities

BOUTONNIÈRE DEFORMITY (FIGURE 3–1; Cailliet, 1982)

MECHANISM

- Weakness or rupture of the terminal portion of the extensor hood (tendon or central slip) at the PIP joint, which holds the lateral bands in place
- Initially caused by PIP joint synovitis
- The lateral bands of the extensor hood slip downward (sublux) from above the axis of the PIP joint to below the axis, turning them into flexors at the PIP joint.
- The PIP then protrudes through the split tendon as if it were a buttonhole (Boutonnière = "buttonhole").
- The distal phalanx hyperextends.

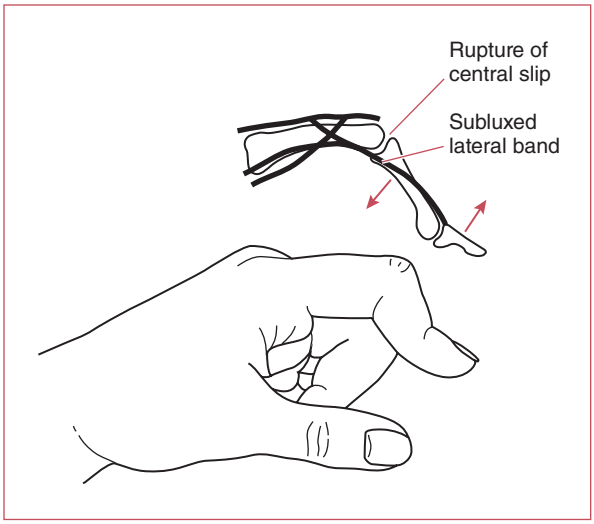

FIGURE 3–1 Boutonnière deformity.

RESULT

- MCP hyperextension
- PIP flexion
- DIP hyperextension

Note: Positioning of the finger as if you were buttoning a button (Boutonnière = "buttonhole").

TREATMENT

- Boutonnière ring splint

SWAN NECK DEFORMITY (FIGURE 3–2; Cailliet, 1982)

MECHANISM

- Common in patients with RA
- Unlike a Boutonnière deformity, a swan neck deformity may be due to synovitis at the MCP, PIP, or DIP (rare) joint.
- Flexor tenosynovitis → MCP flexion contracture
- Contracture of the intrinsic (lumbricals, interosseous) → PIP hyperextension
- Contracture of deep finger flexor muscles and tendons → DIP flexion

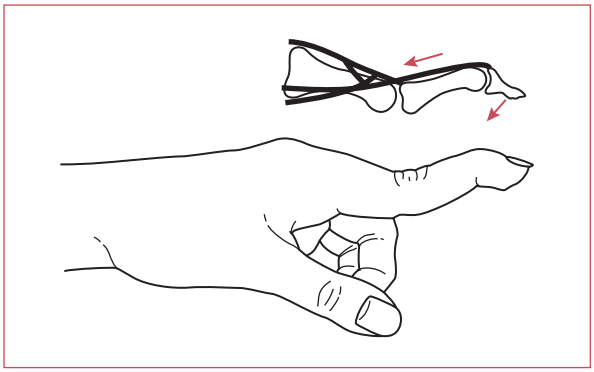

FIGURE 3–2 Swan neck deformity.

RESULT
- MCP flexion contracture
- PIP hyperextension
- DIP flexion

TREATMENT
- Swan neck ring splint orthosis

ULNAR DEVIATION OF THE FINGERS (Cailliet, 1982)

MECHANISM
- Synovitis and weakening of the extensor carpi ulnaris, ulnar, and radial collateral ligaments
- Results in radial wrist deviation; increases the torque of the stronger ulnar finger flexors
- Flexor/extensor mismatch causes *ulnar* deviation of the fingers as the patient tries to extend the joint.

RESULT
- Ulnar deviation is due to the pull of the long finger flexors.
- Radial deviation of the wrist

TREATMENT
- Ulnar deviation splint

FLEXOR TENOSYNOVITIS
- Diffuse swelling of the extensor and flexor tendon sheaths
- One of the most common manifestations of the hands in RA
- Can be a major cause of hand pain and weakness
- 🕮 Early RA may be confused with de Quervain's disease.

INSTABILITY OF THE CARPAL BONES

MECHANISM
- Ligament laxity
- Carpal bone erosions
- Radial deviation of the wrist
- Ulnar styloid rotates dorsally
- Carpal bones rotate
  - Proximal row: Volar direction
  - Distal row: Dorsal direction

RESULT
- The carpal bones rotate in a zigzag pattern.

De Quervain's Tenosynovitis
- Tenosynovitis of the EPB and APL tendons
- Thickening of the tendon sheath results in tenosynovitis and inflammation
- Clinically presents with pain over the radial wrist (EPB and APL tendons)

Test: Finkelstein's test (Figure 3–3)
- Full flexion of the thumb into the palm followed by ulnar deviation of the wrist will produce pain and is diagnostic of de Quervain's tenosynovitis.

APL, abductor pollicis longus; EPB, extensor pollicis brevis.

FIGURE 3–3 Finkelstein's test: Full flexion of the thumb into the palm will produce pain when the wrist is deviated in the ulnar direction. *Source:* From Snider RK, ed. *Essentials of Musculoskeletal Care*. American Academy of Orthopaedic Surgeons; 1997, with permission.

FLOATING ULNAR HEAD ("PIANO KEY SIGN"—THINK OF THE BLACK KEYS)

MECHANISM

- Synovitis at the ulnar styloid leads to rupture or destruction of the ulnar collateral ligament, which results in laxity of the dorsal radioulnar joint (DRUJ).

RESULT

- The ulnar head "floats up" dorsally in the wrist and is easily compressible.
- A positive "piano key sign" assesses the stability of the DRUJ and is positive when the ulnar head is pressed down and springs back up into position once pressure on it is released—think of pressing on piano keys.

RESORPTIVE ARTHROPATHY

MECHANISM

- Digits are shortened and phalanges appear retracted with skin folds.
- Possible mechanism via osteoclastogenesis and osteoclastic bone resorption (Firestein et al., 2008)

RESULT

- Telescoping appearance of the digits
- Most serious arthritic involvement

PSEUDOBENEDICTION SIGN

MECHANISM

- Stretched radioulnar ligaments allow the ulna to drift upward.
- Extensor tendons of the fourth and fifth digit are subject to abrasion from rubbing on the sharp, elevated ulnar styloid and can rupture.

RESULT

- Extensor tendon rupture
- Inability to fully extend the fourth and fifth digits

Shoulder Deformities

- Glenohumeral (GH) arthritis:
  - Limitation of GH internal rotation is an early finding.
- Effusions can occur; decreased range of motion (ROM) may lead to adhesive capsulitis.
- Rotator cuff injuries:
  - Superior subluxations, tears, or fragmenting of tendons secondary to erosion of the greater tuberosity.

Elbow Deformities

- Subcutaneous nodules
- Olecranon bursitis
- Loss of full elbow extension is an early problem and may lead to flexion deformities.
- Ulnar neuropathies

Cervical Spine Instability

- 🕮 A-A joint subluxations → most common are anterior subluxations.
- **Causes in RA:**
  - Tenosynovitis of the transverse ligament of C1 can result in rupture of the ligament and cause subluxation or instability at the A-A joint.
  - Odontoid or atlas erosion
  - Basilar invagination may occur.
  - 🕮 With cervical flexion, the A-A space normally should not increase significantly. Any space larger than 2.5 to 3 mm is considered abnormal (Martel, 1961; Park et al., 1979).

- Instability of the C1 to C2 articulation can cause pain and myelopathy.
- Pre-op C-spine flexion-extension x-rays are recommended in RA patients prior to surgery to ensure there is no cervical instability.

Lower Extremity Deformities

HIP DEFORMITIES

- Occur in about 50% of patients with RA (Duthie & Harris, 1969)
- Symmetric involvement
- Protrusio acetabuli can occur: Deepening of the acetabulum with medial migration of the femoral head
- Accompanied by hip arthritis, usually due to RA

KNEE DEFORMITIES

- Symmetric joint involvement common
- Loss of full knee extension that may lead to flexion contractures
- Quadriceps atrophy leading to increased amount of force though the patella
- Force leads to increased intra-articular pressure in the knee joint, causing the synovial fluid to drip into the popliteal space (i.e., popliteal or Baker's cyst).

ANKLE DEFORMITIES

- Ligament weakness leading to hindfoot pronation
- Tarsal tunnel syndrome:
  - Synovial inflammation can lead to compression of the posterior tibial nerve.

FOOT DEFORMITIES

- Hammer toe deformities:
  - Hyperextension of the MTP and DIP joints with flexion of the PIP joint
- Claw toe deformities:
  - Hyperextension at the MTP joint and flexion of the PIP and DIP joints
  - Pain on the metatarsal heads on weight-bearing
- Rocker bottom foot:
  - Tarsometatarsal joints are inflamed and may sublux, causing a rocker bottom deformity of the foot.
- Morton's neuroma:
  - Forefoot pain due to nerve compression when standing or walking, most commonly found between the third and fourth metatarsal heads, ultimately leading to palpable soft tissue mass on palpation
- Hallux valgus deformity:
  - Lateral deviation of the toes

EXTRA-ARTICULAR MANIFESTATIONS OF RHEUMATOID ARTHRITIS

- It is important to remember that RA is a systemic disease.
- Extra-articular manifestations are more common in patients with the following findings:
  - RF (+)
  - Rheumatoid nodules
  - Severe articular disease
  - MHC class HLA-DRB1 alleles

Constitutional

- Malaise or fatigue

Skin

- Subcutaneous rheumatoid nodules:
  - Present in 50% of RA patients
  - Form subcutaneously, in bursae and along tendon sheaths
  - Typically located over pressure points
  - Extensor surface of the forearm (olecranon)
  - Can occur singly or aggregate in clusters
  - Methotrexate may enhance or accelerate the development of rheumatoid nodules.
- **Vasculitic lesions:**
  - Leukocytoclastic vasculitis and palpable purpura
  - Lower extremity ulcers

> 📖 **Subcutaneous Nodules Are Seen in:**
> - Rheumatoid arthritis
> - Gout

Ocular

- Keratoconjunctivitis sicca (dry eye syndrome)
- Episcleritis → benign, self-limited
- Scleritis → severe inflammation may erode through the sclera into the choroid, causing scleromalacia perforans.
- Peripheral ulcerative keratitis → immune-mediated vasculitis in the terminal vessels at the periphery of the cornea causes crescent-shaped ulcer in the peripheral cornea.

Pulmonary

- Interstitial lung disease (ILD):
  - Asymptomatic nodules
  - Interstitial fibrosis
- Pleurisy
- Inflammation of the cricoarytenoid joint → dysphagia, dysphonia

> **Caplan's Syndrome**
> - Intrapulmonary nodules—histologically similar to rheumatoid nodules
> - RF (+)
> - Associated with RA and pneumoconiosis in coal workers
> - Granulomatous response to silica dust
>
> RA, rheumatoid arthritis; RF, rheumatoid factor.

Cardiac

- 📖 Pericarditis: Classic findings include chest pain, pericardial friction rub, and EKG abnormalities (diffuse ST elevations).
  - May lead to constrictive pericarditis with right-sided heart failure
  - May be found in about half of RA patients
  - Rarely symptomatic
- Valvular heart disease

Gastrointestinal

- Xerostomia—dryness of the mouth secondary to decreased salivary secretion
- Gastritis and peptic ulcer disease (PUD) associated with nonsteroidal anti-inflammatory drug (NSAID) use—not directly linked to disease

Renal

- Glomerulonephritis is rare.
- May see renal involvement if amyloidosis develops

Neurologic

- Cervical spine (see the previous section)
  - Most common at C1 to C2: Erosions of the transverse ligament as well as apical and alar ligaments or the dens itself, which can lead to cervical subluxation/instability and myelomalacia

  - Cervical myelomalacia:
    - Gradual onset of bilateral sensory paresthesia and motor weakness in the upper extremity (UE), lower extremity (LE), or both. Can result in dexterity and balance deficits as well as neurogenic bower and bladder.
    - Neurologic exam findings may include → positive Babinski's and/or Hoffman's signs, hyperreflexia, and myelopathic gait.
- Entrapment neuropathies:
  - This is secondary to fluctuation in synovial inflammation and joint postures.
- Mononeuritis multiplex—inflammatory—not due to compression

Hematologic

- Hypochromic microcytic anemia
- Thrombocytosis
- Lymphadenopathy (risk for non-Hodgkin's lymphoma)
- Felty's syndrome

Felty's Syndrome

"She felt her spleen."

- 🕮 Classic triad of RA, splenomegaly, and leukopenia
- Seen in seropositive RA, usually with nodules
- Occurs in the fifth or seventh decades with RA >10 years
- Women comprise two-thirds of cases
- Often associated with leg ulcers

RA, rheumatoid arthritis.

TREATMENT OF RHEUMATOID ARTHRITIS (TABLE 3–1; Berkow & Elliott, 1995; Hicks & Sutin, 1988)

Education

- Joint protection education
- Home exercise program
- Studies have shown that patient education combined with strengthening exercises led to less pain than in controls (Stitik et al., 2010).

TABLE 3–1 Treatment Options for RA

DISEASE STAGE	POOR PROGNOSTIC FEATURES	TREATMENT	MEDICATIONS
Mild	(–)	1. Education 2. PT/OT 3. Compliance	1. NSAIDs, salicylates 2. DMARDs: – Hydroxychloroquine – Sulfasalazine – Methotrexate
Moderately severe	(+)	1. Education 2. PT/OT 3. Compliance	1. NSAIDs, salicylates 2. DMARDs, methotrexate 3. Corticosteroids: Typically <10 mg daily prednisone 4. Biological agents
Severe	(+)	1. Education 2. PT/OT 3. Compliance	1. NSAIDs, salicylates 2. DMARDs—chronic use: – Weekly oral or parenteral methotrexate – May add a second DMARD 3. Corticosteroids: Typically prednisone <10 mg daily 4. Biological agents

DMARD, disease-modifying antirheumatic drug; NSAID, nonsteroidal anti-inflammatory drug; OT, occupational therapy; PT, physical therapy.

Source: Adapted From Verhoeven AC, Boers M, Tugwell P. Combination therapy in rheumatoid arthritis: updated systematic review. *Br J Radiol*. 1998;37:612–619. doi:10.1093/rheumatology/37.6.612, with permission.

Relative Rest

- Required for acutely inflamed joint

Exercise

- In acute disease with severely inflamed joints, splinting is used to immobilize the joint, with twice daily, full and slow passive ROM to prevent soft tissue contracture.
- 🕮 Mild disease (moderate synovitis) requires isometric program.
- **Isometric exercise:**
  - Causes the least amount of periarticular bone destruction and joint inflammation/pain, especially during an acute flare
  - Restores and maintains strength
  - Generates maximal muscle tension with minimal work, fatigue, and stress
  - Isotonic and isokinetic exercise may exacerbate the flare and should be avoided.

Modalities

- Superficial moist heat:
  - Should not be used in acutely inflamed joints
  - Increases collagenase enzyme activity that causes increased joint destruction
  - Heat penetration to depth of 1 cm can decrease pain and increase collagen extensibility
- Other superficial heating modalities include paraffin and fluidotherapy.
- Cryotherapy:
  - Pain relief in an acutely inflamed joint
  - Decreases the pain indicators of inflammation

Orthoses

- Splinting of the wrist in RA will most likely help decrease pain and inflammation (Stenger et al., 1998).
- The MCP is thought to be the primary site of RA, and inflammation can lead to weakening of joint supporting structures. The major function of an orthosis used to combat early deformity in RA is to prevent MCP flexion.

Indications for Orthotic Use
- Decrease pain and inflammation
- Reduce weight through joint
- Decrease joint motion—stabilization
- Joint rest

- Remember: There is no evidence that splinting will stop these deformities, but stabilizing the MCP joint accompanied with exercise may help prevent or slow the progression.
- Wrist-hand orthosis contributes to a decrease in pain and is used mainly for strength maneuvers rather than dexterity.
- Does not deter the progression of the disease

Pharmacotherapy in Rheumatoid Arthritis

- One of the main goals of pharmacotherapy in RA is to reduce systemic inflammation to reduce/prevent systemic sequelae, including joint erosion and deformities.
- Commonly used medications include nondisease modifying antirheumatic drugs (non-DMARDs such as NSAIDs, glucocorticoids) and DMARDs, which include nonbiological and biological types.
  - Non-DMARDs: The use of NSAIDs and corticosteroids (oral and intra-articular) is limited due to their efficacy. NSAIDs are typically used as monotherapy only for well-controlled, mild RA. Corticosteroids help reduce acute inflammation but are less efficacious in the long term.
  - DMARDs are the mainstay of pharmacotherapy for RA. They are typically added early on and are frequently used in combination with other DMARDs.

NONDISEASE MODIFYING ANTIRHEUMATIC DRUGS

- Aspirin (ASA), NSAIDs:
  - *Side effects*: Gastrointestinal (GI) ulceration and bleeding, renal insufficiency, hepatotoxicity, hypertension
  - 🕮 Therapeutic range for ASA is 15 to 25 mg/dL. Toxic >30 mg/dL.
- Corticosteroids:
  - *Side effects*: Hyperglycemia, inhibits immune response, osteoporosis, PUD, emotional lability

NONBIOLOGICAL DISEASE MODIFYING ANTIRHEUMATIC DRUG AGENTS

- 🕮 See Table 3–2.

TABLE 3–2 Nonbiological DMARDs Used in RA and Common Side Effects

DMARDS	TOXIC PROFILE	GENERAL DEGREE OF TOXICITY
Hydroxychloroquine	Retinopathy, hyperpigmentation	Safer
Sulfasalazine	Myelosuppression, GI disturbances	Safer
Methotrexate	Stomatitis, myelosuppression, hepatic fibrosis, cirrhosis—requires liver function testing, pulmonary involvement, worsens rheumatoid nodules, teratogenicity	More toxic
Leflunomide	Hepatotoxicity, nausea, diarrhea, hypertension, teratogenicity	More toxic
Cyclosporine	Renal dysfunction, tremor, hirsutism, hypertension, gum dysplasia	More toxic
Gold intramuscular, oral	Myelosuppression, renal → proteinuria Diarrhea (#1, oral), rash (#1, intramuscular)	More toxic
Azathioprine	Myelosuppression, hepatotoxicity, lymphoproliferative disorders	More toxic

DMARD, disease-modifying antirheumatic drug; GI, gastrointestinal; RA, rheumatoid arthritis.
Source: Adapted from Gerber LH, Hicks JE. Surgical and rehabilitation options in the treatment of the rheumatoid arthritis patient resistant to pharmacologic agents. *Rheum Dis Clin North Am*. 1995;21:19–39.

BIOLOGICAL DISEASE MODIFYING ANTIRHEUMATIC DRUG AGENTS

- **Anti-TNF agents:**
  - *Mechanism of action*: Reduce levels of TNF-alpha (etanercept = soluble receptor; infliximab = chimeric antibody; adalimumab = human monoclonal antibody)
  - *Toxicity*: Infection (especially TB and hepatitis B virus [HBV] reactivation), demyelinating disease, induction of autoimmunity, exacerbation of congestive heart failure, possible malignancy (skin cancer), hepatosplenic T-cell lymphoma (HSTCL), rare aggressive and fatal disease seen in patients treated with TNF blockers
  - *Examples*: Etanercept (Enbrel®), infliximab (Remicade®), and adalimumab (Humira®)
- **Co-stimulation modulators:**
  - *Mechanism of action*: Prevent T-cell activation by interfering with antigen-presenting cell interaction with T-cells
  - *Toxicity*: Infection, exacerbation of chronic obstructive pulmonary disease
  - *Example*: Abatacept (Orencia®)

- **Anti-B-cell antibodies:**
  - *Mechanism of action*: Depletes B-cells
  - *Toxicity*: Infection, death, central nervous system (possible progressive multifocal leukoencephalopathy)
  - *Example*: Rituximab
- **IL-1 receptor antagonist:**
  - *Mechanism of action*: Antagonizes IL-1 by binding to IL-1 receptor
  - *Toxicity*: Injection site reactions, infection
  - *Example*: Anakinra (Kineret®)
- **IL-6 antagonist:**
  - *Mechanism of action*: Antagonizes IL-6 by binding to IL-6 receptor
  - *Toxicity*: Transaminase elevation, leukopenia, thrombocytopenia, hyperlipidemia, bowel perforation, infection
  - *Example*: Tocilizumab (Actemra®)
- **Protein kinase inhibitors:**
  - *Mechanism of action*: Inhibits intracellular protein kinases (Janus kinase [JAK])
  - *Toxicity*: Leukopenia, anemia, transaminase elevation, hyperlipidemia, bowel perforation, infection
  - *Example*: Tofacitinib (Xeljanz®)

Surgical Intervention in Rheumatoid Arthritis

- C1–C2 fusion for A-A instability
- Synovectomy: Most commonly with extensor tenosynovitis at the wrist
- Arthroplasty: Knee and hip most common; shoulder and MCP infrequent; elbow rare
- Arthrodesis: Typically for ankle, occasionally for wrist or thumb
- Tendon repairs: Generally unsuccessful; most hand/wrist tendinopathies require tendon transfer

Poor Prognostic Factors in RA

1. Rheumatoid nodules
2. RF (+)
3. x-ray consistent with erosive disease
4. Persistent synovitis
5. Insidious onset
6. Anti-CCP antibodies

CCP, cyclic citrullinated peptide; RA, rheumatoid arthritis; RF, rheumatoid factor.

OSTEOARTHRITIS

- Osteoarthritis (OA) is a nonerosive, noninflammatory progressive disorder of the joints leading to deterioration of the articular cartilage and new bone formation at the joint surfaces and margins.
- OA is a disease of the cartilage initially, not bone.
- OA is a clinical diagnosis.
- Although OA has long been known as a noninflammatory disease, later in the disease inflammatory-induced and stress-induced signaling pathways may be activated in the synovium and cartilage (due to changes in the environment of the joint; Knights et al., 2023).

PREVALENCE

- Most common form of arthritis in adults, and the second most common form of disability in the United States
- Prevalence increases with age: Approximately 70% of population >65 years old has radiographic evidence of OA (Lane, 1997). Of the 32.5 million people in the United States with a diagnosis of OA, 18.7 million people are between the ages of 18 and 64 years (CDC, 2020).

- Increase in occupations with repetitive trauma
- Male:female ratio is equal between ages 45 and 55. After the age of 55, it is more common in women.
- Obesity → OA of the knee is most common, but there is also an increased rate of OA in non-weight-bearing joints, which is thought to be due to increased systemic inflammatory activity in obesity (Nedunchezhiyan et al., 2022).
- Obesity → OA of the knee is most common.

🕮 PATHOLOGY

- Early → Hypercellularity of chondrocytes:
  - Cartilage breakdown: Swelling and loosening of collagen framework
  - Increased proteoglycan synthesis
  - Minimal inflammation
- Later → Cartilage fissuring, pitting, and destruction:
  - Hypocellularity of chondrocytes
  - Inflammation secondary to synovitis
  - Osteophyte spur formation—seen at the joint margins
  - Subchondral bone sclerosis (eburnation)
  - Cyst formation in the juxta-articular bone
- Loss of proteoglycans
- 🕮 Increased water content of OA cartilage leads to damage of the collagen network (increased chondrocytes, collagen, and enzymes).

Commonly Affected Joints in OA
- Primary OA: Knees, MTP, DIP, CMC, hips, and spine (facet and uncovertebral joints)
- Secondary OA: Elbows and shoulders

CMC, carpal metacarpal joint; DIP, distal interphalangeal; MTP, metatarsophalangeal; OA, osteoarthritis.

CLASSIFICATION OF OSTEOARTHRITIS

- Primary OA (idiopathic):
  - Knees, MTP, DIP, carpal metacarpal joint (CMC), hips, and spine primarily involved
- Secondary OA → follows a recognizable underlying cause:
  - Elbows and shoulder involvement
  - Chronic or acute trauma, connective tissue disease (CTD), endocrine or metabolic, infectious, neuropathic, crystal deposition, bone dysplasias
- Erosive inflammatory OA
  - Variant of hand OA characterized by acute-onset pain, swelling, and redness in DIP and PIP joints of the hands, causing joint space narrowing, erosions, and ankylosis.
  - Osteophyte formation and erosions create characteristic "sawtooth" and "gull wing" lesions on x-rays.
- Diffuse idiopathic skeletal hyperostosis (DISH; Snider, 1997):
  - Most commonly affects the thoracic spine but can also affect the lumbar and cervical spines

Diffuse Idiopathic Skeletal Hyperostosis

- Variant form of spinal arthritis typically characterized by the formation of nonmarginal (away from the margins of the vertebral body) syndesmophytes along the anterolateral spinal column and resulting in ossification of spinal ligaments
- Nonmarginal syndesmophytes extending to the length of the ALL on the right side of the anterior spine leading to spinal fusion typically in the thoracic or thoracolumbar spine
- Commonly asymmetric with predilection for the right side of the thoracic spine
- 🕮 Hallmark → ossification spanning four contiguous vertebral bodies (three or more intervertebral discs)
- Ossification of the ALL, separated from vertebral body by radiolucent line
- More prevalent in White males above the age of 45
- Multisystem disorder associated with:
  - DM, obesity, hypertension, coronary artery disease
  - Stiffness in the morning or evening
  - 🕮 Dysphagia with cervical involvement

🕮 NOT associated with sacroiliitis, apophyseal joint ankylosis, or HLA-B27 positivity (distinguishes from ankylosing spondylitis; Snider, 1997)

ALL, anterior longitudinal ligament; DM, diabetes mellitus; HLA-B27, human leukocyte antigen B27.

SIGNS AND SYMPTOMS OF OSTEOARTHRITIS

- **Symptoms:**
  - Dull aching pain increased with activity, relieved by rest
  - Later pain occurs at rest
  - Joint stiffness for <30 minutes; becomes worse as the day goes on
  - Joint giving away
  - Articular gelling → stiffness after immobility lasting short periods and dissipating after brief period of movement
  - Crepitus on ROM
- **Signs:**
  - Mono- or pauciarticular; shows no obvious joint pattern
  - Localized tenderness of joints
  - Pain and crepitus of involved joints
  - Enlargement of the joint → changes in the cartilage and bone secondary to proliferation of synovial fluid and synovitis

SPECIFIC JOINT INVOLVEMENT

- Heberden's nodes → osteophytosis (bone spur formation) at the DIP joints
- Bouchard's nodes → PIP joints
- First CMC joint
- Knee—narrowing of medial compartment
- Hips—narrowing of superior lateral joint space compartment, loss of ROM
- Acromioclavicular (AC) joint
- First MTP joint
- Spine:
  - Spondylosis
  - Involvement of the intervertebral fibrocartilaginous disc and vertebrae
  - DISH (Snider, 1997; see section on DISH)

RADIOGRAPHIC FINDINGS (TABLE 3–3)

- Asymmetric narrowing of the joint space:
  - Knee—medial compartment narrowing most common
  - Hip—superolateral joint space narrowing more common
- Subchondral bony sclerosis (eburnation)—new bone formation (white appearance)
- Osteophyte formation
- Osseous cysts—can lead to microfractures and bony collapse
- Loose bodies, joint effusion
- Not associated with osteoporosis/osteopenia (no bone washout)
- Joint involvement:
  - First CMC joint
  - PIP—Bouchard's nodes; DIP—Heberden's nodes
  - Large joints—knee and hip
  - Luschka's (uncovertebral) joints—uncinate process on the superior/lateral aspect of the cervical vertebral bodies (C3–C7), making them concave

TREATMENT

- Patient education:
  - Weight loss
  - Activity modification
- Physical therapy (PT)/occupational therapy (OT):
  - ROM, strengthening exercises
  - Assistive devices
  - Joint protection and energy conservation
  - Tai Chi strongly recommended, and cognitive behavioral therapy and yoga conditionally recommended, for knee OA
- 🕮 Pharmacotherapy:
  - Acetaminophen (initial treatment)
  - NSAIDs (used for pain once inflammation ensues)
  - Narcotics—rare
  - Oral steroids anecdotally effective in inflammatory/erosive OA
  - Limited recommendations for acupuncture
- Osteotomy: In patients with unicompartmental OA of the knee with varus or valgus malalignment, high tibial or distal femoral osteotomy can be performed in order to transfer the load bearing onto the healthy compartment of the knee. This has been shown to improve knee function and reduce pain; however, it has not been directly compared with conservative measures (Loia et al., 2016).
  - Indications for osteotomy (Loia et al., 2016):
    - Age: Unsuccessful surgery risk is five times higher for patients older than 56.
    - Body mass index (BMI): Ideal BMI between 25 and 27.5
    - ROM: >120 degrees, flexion contracture <5 degrees
- Unicompartmental knee replacement:
  - In patients with advanced OA limited to a single compartment
  - During this procedure, the damaged compartment is replaced with metal or plaster. The healthy parts of the knee, bone, and cartilage, as well as ligaments, are preserved.
- Intra-articular injections:
  - Intra-articular steroid injections may be beneficial in acute flares.
  - Hyaluronic acid viscosupplementation:
    - Although still used clinically to treat mostly knee OA, it is *not recommended* routinely when treating symptomatic knee OA (American Academy of Orthopaedic Surgeons [AAOS], 2021).
    - Conditionally recommended *against* in knee OA or first CMC joint OA (Kolasinski et al., 2020)
    - Strongly recommended *against* in hip OA (Kolasinski et al., 2020)
- The Framingham Study showed that women who had a decrease in BMI of 2 units (weight loss of 5.1 kg or 11.22 lb. over 10 years) decreased the odds for developing symptomatic knee OA by over

50%. Additionally, it has been shown that patients with existing OA of weight-bearing joints had a considerable improvement in pain, function, and knee ROM during stance and swing phases after losing an average of 100 lb. over a 12- to 18-month period (Felson et al., 1992; Stitik et al., 2010).

- Flexibility, strength, and proprioception are maximized to reduce joint stresses and maximize movement and alignment.
- Patellofemoral taping techniques have been used to relieve knee pain in OA and improve function.
- Lower limb braces can be prescribed for patients with OA in order to inhibit multiplane joint activity and relieve pain associated with weight-bearing. Supporting the affected joint, reducing the muscular force needed to stabilize it, and redirecting axial loads all contribute to conservation of the joint and preventing further damage.
- Knee bracing can be used for medial compartment OA that utilizes a three-point force system across the coronal plane, forcing the tibia in a valgus direction, thereby unloading the medial compartment. This has been shown to reduce external varus movement at the knee during initial contact and loading response phases (Stitik et al., 2010).
  - A simple knee sleeve may be used for mild OA without angular deformity.
  - Wedged shoe soles/insoles may be used to counter knee joint compressive forces, which work to reduce the axial load on the medial joint surface.
- A thumb orthosis for first CMC OA would place the thumb in metacarpal abduction, align the metacarpal on the trapezium, flex the metacarpophalangeal (MP) joint, and extend the IP joint, which decreases pain and increases function, conserving the joint (Beasley, 2012).

COMPARISON OF OSTEOARTHRITIS VERSUS RHEUMATOID ARTHRITIS

OA Versus RA Erosive X-Ray Changes (Table 3–3)
OA → No erosive changes seen on x-ray
RA → Marginal or central erosions seen on x-ray

OA, osteoarthritis; RA, rheumatoid arthritis.

TABLE 3–3 Rheumatoid Arthritis Versus Osteoarthritis Comparison

RHEUMATOID ARTHRITIS	OSTEOARTHRITIS
RADIOGRAPHIC FINDINGS	
• Uniform joint space narrowing—hips, knees, and so forth • Symmetrical joint involvement • Marginal bone erosions • Juxta-articular osteopenia—bone washout • Ulnar deviation of phalanges • Radial deviation of the radiocarpal joint (carpals and metacarpals) • Erosion of the ulnar styloid • Atlantoaxial separation (>2.5–3 mm) subluxation • Small joint involvement—MCP, PIP, carpal	• Asymmetric narrowing of the joint space: – Knee—medial joint space narrowing more common – Hip—superior lateral joint space narrowing more common • Joint involvement does not have to be symmetric • No erosive changes seen on x-ray • No osteoporosis/osteopenia (bone washout) • Subchondral bony sclerosis—new bone formation with white appearance • Osteophytosis • Osseous cysts—microfractures may cause bony collapse • Loose bodies • Joint involvement; first CMC, DIP, large joints—knee and hip
EXTREMITY INVOLVEMENT	
• Wrist • MCP • PIP • Ankle joint • Talonavicular joint • MTP	• First CMC • PIP • DIP • MTP

CMC, carpometacarpal joint; DIP, distal interphalangeal; MCP, metacarpophalangeal; MTP, metatarsophalangeal; PIP, proximal interphalangeal.

JUVENILE IDIOPATHIC ARTHRITIS (FORMERLY JUVENILE RHEUMATOID ARTHRITIS; SEE TABLE 3–4)

(Also Refer to Juvenile Idiopathic Arthritis [JIA] in Chapter 10, Pediatric Rehabilitation)

- Formerly known as juvenile rheumatoid arthritis (JRA), juvenile idiopathic arthritis (JIA) is the most common form of childhood chronic inflammatory rheumatic disease (Minden, 2023).
- Cause unknown; is a collection of inflammatory arthritides (Ringold et al., 2019)
- A diagnosis of exclusion that is identified clinically
- Chronic arthritic disease in children; may persist into adulthood
- All associated with decreased health-related quality of life and increased permanent joint damage risk (Ringold et al., 2019)
- The International League of Associations for Rheumatology (ILAR) has established seven clinical categories of JIA, including systemic, RF-negative polyarticular, RF-positive polyarticular, oligoarticular, psoriatic, enthesis-related, and undifferentiated (Table 3–4).

International League of Associations for Rheumatology (ILAR) Classification Criteria for JIA

- Onset <16 years of age
- Persistent arthritis in one or more joints for at least 6 weeks
- Exclusion of other types of childhood arthritis—rheumatic fever, infection, SLE, vasculitis, leukemic arthritis, and so forth

JIA, juvenile idiopathic arthritis; SLE, systemic lupus erythematosus.

- While the ILAR has defined seven categories of JIA, the ACR uses clinical phenotypes of JIA (instead of the ILAR categories) to base treatment guidelines (Ringold et al., 2019). The ACR has published treatment guidelines for the following clinical phenotypes: nonsystemic polyarthritis, sacroiliitis, enthesitis, oligoarthritis, temporomandibular joint arthritis, systemic JIA, and JIA-associated uveitis (Angeles-Han et al., 2019; Onel et al., 2022a, 2022b; Ringold et al., 2019).

TABLE 3–4 International League of Associations for Rheumatology Classification (ILAR) of Juvenile Idiopathic Arthritis (JIA)

JIA CATEGORY	DEFINITION
Systemic arthritis	Arthritis in ≥1 joint with daily fever of ≥2 weeks duration plus ≥1 of: 1. Evanescent, erythematous rash 2. Generalized lymph node enlargement 3. Hepatomegaly and/or splenomegaly 4. Serositis Exclusions: A, B, C, D
Oligoarthritis • Persistent • Extended	Arthritis affecting 1–4 joints during the first 6 months of disease • Affecting ≤4 joints throughout the disease course • Affecting a total of >4 joints after the first 6 months of disease Exclusions: A, B, C, D, E

(*continued*)

TABLE 3–4 International League of Associations for Rheumatology Classification (ILAR) of Juvenile Idiopathic Arthritis (JIA) (*continued*)

JIA CATEGORY	DEFINITION
Rheumatoid factor negative polyarthritis	Arthritis affecting ≥5 joints during the first 6 months of disease; a test for rheumatoid factor is negative Exclusions: A, B, C, D, E
Rheumatoid factor positive polyarthritis	Arthritis affecting ≥5 joints during the first 6 months of disease; ≥2 tests for rheumatoid factor ≥3 months apart are positive in the first 6 months of disease Exclusions: A, B, C, E
Psoriatic arthritis	Arthritis and psoriasis OR arthritis plus ≥2 of: 1. Dactylitis 2. Nail pitting or onycholysis 3. Psoriasis in a first-degree relative Exclusions: B, C, D, E
Enthesitis-related arthritis	Arthritis and enthesitis OR arthritis OR arthritis plus ≥2 of: 1. Sacroiliac joint tenderness and/or inflammatory lumbosacral pain 2. Presence of HLA-B27 antigen 3. Acute anterior uveitis 4. History of spondyloarthritis or acute anterior uveitis in a first-degree relative Exclusions: A, D, C
Undifferentiated arthritis	Arthritis fulfilling criteria in no category or in ≥2 categories

Exclusions: A, psoriasis or a history of psoriasis in the patient or a first-degree relative; B, arthritis in an HLA-B27 positive male beginning after the sixth birthday; C, spondyloarthritis, enthesitis-related arthritis or acute anterior uveitis OR a history of these disorders in a first-degree relative; D, presence of IgM rheumatoid factor on ≥2 occasions ≥3 months apart; E, presence of a systemic JIA in the patient.
HLA, human leukocyte antigen; IgM, immunoglobulin M.
Source: From Standardization of Uveitis Nomenclature (SUN) Working Group. Classification Criteria for Juvenile Idiopathic Arthritis-Associated Chronic Anterior Uveitis. *Am J Ophthalmol*. 2021;228:192–197. doi:10.1016/j.ajo.2021.03.055. Epub 2021 May 11. PMID: 33845021; PMCID: PMC8594759.

CLINICAL SUBTYPES OF JUVENILE IDIOPATHIC ARTHRITIS

Other Characteristics of Juvenile Idiopathic Arthritis Categories

1. **Systemic arthritis (JIA)—"Still's disease":**
   - Accounts for approximately 5% of JIA patients
   - Onset peak—1 to 6 years old, male = female
   - Multisystem involvement may also include:
     - Growth delay
     - Osteoporosis, osteopenia
     - Pericarditis
     - Pleuritis
     - Anemia
     - Leukocytosis
     - Elevated acute phase reactants
   - RF (+) <2%
   - Uveitis rare
2. **Polyarticular JIA:**
   - Polyarticular JIA: Five or more joints involved during the first 6 months
   - Mild systemic manifestations
   - Females >> males, onset usually >8 years old

- Gradual onset of swelling, stiffness involving the cervical spine and hips
- Growth retardation due to early closure of the epiphyseal plates in the long bones (limb length discrepancy) and the jaw (micrognathia)
- RF (–): 90% to 95%
- RF (+): 5% to 10%

RF (+) POLYARTICULAR (5%–10%)	RF (–) POLYARTICULAR (90%–95%)
• Females >10 years old • Erosive and chronic • Unremitting: This group has the worst prognosis if disease is unremitting • Uveitis does not occur • Subcutaneous nodules	• 25% males <6 years old

RF, rheumatoid factor.

3. **Oligoarticular JIA:**
   - Most common form of JIA in White populations from North America and Europe (Minden, 2023)
     - Female >> male
     - Female onset between ages 1 and 5
     - Male onset in late school age or adolescence
   - Oligoarticular JIA: Four or fewer joints affected during the first 6 months
   - 🕮 Knee is the most common joint involved, followed by ankle, wrist, and then elbows.
   - Hips usually spared (–) sacroiliitis
   - Limb length discrepancy may develop (affected side longer) due to increased blood flow to growing bone in inflammatory state.
   - Two distinct types:
     - Persistent: No additional joint involvement after the first 6 months, remains <4 joints
     - Extended: Additional joint involvement after the first 6 months, such that disease becomes polyarticular (five or more joints)
   - (+) HLA-B27
   - RF (+) <2% (Klippel, 1997)
   - Antinuclear antibody (ANA) (+) patients have greater risk of eye involvement.
     - Chronic iridocyclitis (often asymptomatic)—leads to cataracts, glaucoma, or blindness if not discovered—requires frequent ophthalmology surveillance
     - Key points about oligoarticular JIA (see the following box).

🕮 **Key Points**
- ANA (+), RF (–)
- HLA-B27 (+)

ANA, antinuclear antibody; HLA, human leukocyte antigen; RF, rheumatoid factor.

🕮 **Ophthalmology Referral Is Mandatory:**
- At risk for developing uveitis and iridocyclitis, which may be asymptomatic
- Slit lamp exam is required four times per year, for 4–5 years; no erosions

MANAGEMENT OF JUVENILE IDIOPATHIC ARTHRITIS

- 70% of children with JIA improve without serious disability (worst prognosis if polyarticular with unremitting disease).
- 10% of children suffer severe functional disability.

- A healthy, well-balanced, age-appropriate diet is recommended, along with immunizations and shared decision-making with patients and caregivers (Onel et al., 2022a, 2022b).
- Use of screening x-rays (prior to advanced imaging) to identify active synovitis or enthesitis is strongly recommended *against* (Onel et al., 2022a, 2022b).
- Poor prognosis indicators are specific to each clinical phenotype and may include:
  - RF (+) (if history of arthritis of five or more joints)
  - Bony erosions or joint space narrowing on x-ray
  - Arthritis in the hip or cervical spine
- Therapeutic goals (Onel et al., 2022a, 2022b):
  - Include physical and occupational therapy (evaluate case by case)
  - Relieving symptoms and limiting functional impairments; improving participation in activities of daily living and family and community activities
  - Maintaining and improving joint ROM
  - Improve strength and endurance
  - Joint protection, prevent injury
  - Family training
- Pharmacotherapy (Amgees-Han et al., 2019; Onel et al., 2022a, 2022b; Ringold et al., 2019):
  - While the ACR's published treatment guidelines are for the clinical phenotypes, most of the recommendations are conditional and have a low quality of supporting evidence. The onus is placed on the clinician, in conjunction with the patient and caregivers, to determine the correct treatment regimen.
  - Specific laboratory monitoring for medications may be required.
  - 🕮 Salicylates should be avoided if possible because of the risk of Reyes' syndrome.
  - Medications that may be used in different clinical scenarios include:
    - NSAIDs with food and GI protection. Tolmetin, naproxen, and ibuprofen are approved (naproxen and ibuprofen available as liquid).
    - Corticosteroids (preferably intra-articular such as triamcinolone hexacetonide)
    - Conventional DMARDs: Methotrexate, sulfasalazine, and leflunomide. Methotrexate is more commonly preferred over other options in oligoarthritic and polyarticular types.
    - Biological DMARDs: TNF inhibitors, IL-1-RA (anakinra), IL-6-RA (tocilizumab)
- Surgical correction of deformities if severe and disease is advanced (such as arthrodesis, synovectomy, and arthroplasty)

JUVENILE SPONDYLOARTHROPATHIES

GENERAL

- Usually takes many years to diagnose
- Symptoms before or at the age of 16
- HLA-B27 (+) associated disorders (most common subtype; majority of heritable ankylosing spondylitis), although is not necessary for diagnosis (Gmuca & Weiss, 2015)

CLASSIFICATION

1. Differentiated forms include juvenile psoriatic arthritis, enthesis-related arthritis, juvenile ankylosing spondylitis (RF [−], ANA [−]), reactive arthritis, and arthropathies associated with inflammatory bowel disease (IBD).
   - Since these diseases take many years to fully evolve, it is not uncommon for children to have symptoms but not satisfy full classification criteria.

- Because of this, it is suggested that spondyloarthropathies in children include another syndrome, seronegative enthesopathy and arthropathy (SEA).
- SEA syndrome:
  - RF (–) and ANA (–)
  - Characterized by enthesitis, arthritis or arthralgia, axial skeletal involvement, and genetic predisposition (HLA-B27)
  - Extra-articular manifestations may include bowel inflammation, psoriasis, cardiac disease, and acute anterior uveitis.

TABLE 3–5 Key Points of Juvenile Arthritides: JIA Versus Juvenile Spondyloarthropathies

JUVENILE IDIOPATHIC ARTHRITIS			
SYSTEMIC MULTISYSTEMIC INVOLVEMENT	**POLYARTICULAR (5+ JOINTS INVOLVED)**	**OLIGOARTICULAR (1–4 JOINTS INVOLVED)**	**JUVENILE SPONDYLOARTHROPATHIES**
RF (–): ~98% Still's disease High fever Rheum. rash Lymphadenopathy Hepatosplenomegaly Anemia May develop into MAS, a severe, potentially fatal inflammatory reaction; markers of inflammation (leukocytosis, sedimentation rate elevation) commonly present in systemic JIA flares will reverse and drop acutely in MAS.	RF (–): 90%–95% • No extra-articular manifestations of systemic-onset disease • Gradual onset of: – Swelling – Stiffness – + cervical spine – + hip – +TMJ (can lead to micrognathia) • Growth retardation • Early closure of epiphyseal plates Note: This group has the worst prognosis when disease is unremitting.	RF (–): >98% • 1–4 joint involvement • Few systemic effects • Chronic iridocyclitis: <6 years occurs in 20%–40%; more frequently in females with ANA (+) • Must have ophthalmology exam four times/first year and annually × 4 years – (+) HLA-B27 – (–) Bony erosions on x-ray	RF (–) HLA-B27 (+) in enthesitis-related arthritis • Juvenile psoriatic arthritis • Enthesis-related arthritis • Juvenile ankylosing spondylitis • Reactive arthritis • Arthropathies associated with inflammatory bowel disease • Overall poorer prognosis compared with other JIA categories (Gmuca & Weiss, 2015) Presents like adult spondyloarthropathies (see later) • SEA syndrome: – RF (–) – ANA (–) – Enthesitis/arthritis/arthralgia – May have uveitis (painful and acute)

INFLAMMATORY ARTHRITIS	NONINFLAMMATORY ARTHRITIS
• Increase in WBC and ESR • Acute painful onset • Erythema, warmth, and tenderness • CTD—SLE, polymyositis/dermatomyositis, PSS, RA • Crystal—gout and pseudogout • Infectious • Seronegative spondyloarthropathies	• Degenerative joint disease—OA, AVN • Traumatic • Joint tumors • Hemophilia • Metabolic—hemochromatosis, alkaptonuria, rheumatic fever, Wilson's disease

ANA, antinuclear antibody; AVN, avascular necrosis; CTD, connective tissue disease; ESR, erythrocyte sedimentation rate; HLA, human leukocyte antigen; JIA, juvenile idiopathic arthritis; MAS, macrophage activation syndrome; OA, osteoarthritis; PSS, progressive systemic sclerosis; RA, rheumatoid arthritis; RF, rheumatoid factor; SEA, seronegative enthesopathy and arthropathy; SLE, systemic lupus erythematosus; TMJ, temporomandibular joints; WBC, white blood cells.

CRYSTAL-INDUCED SYNOVITIS (TABLE 3–6)

TABLE 3–6 Crystal-Induced Synovitis

	GOUT	PSEUDOGOUT
🕮 Crystal	• Monosodium urate crystals • Acute synovitis in the synovial membrane and joint cavity	• "Articular chondrocalcinosis" • CPPD crystals • Hyaline cartilage and fibrocartilage joints
Aspirate: Microscopic	🕮 *Negative* birefringence (moderate to severe inflammation, WBC 15,000–20,000—neutrophils)	🕮 *Positive* birefringence
Epidemiology	Male >> female Age—30–50 years	Male > female Age—30–50 years
Sequelae	• Gouty arthritis • Acute recurrent attacks • Chronic tophaceous arthritis • Uric acid calculi • Urate nephropathy	• Acute pseudogout • Inflammatory host response to CPPD crystals shed from the cartilaginous tissues to the synovial cavity 🕮 Pseudogout may have associations with: Hypothyroidism, Hyperparathyroidism, Hemochromatosis; Amyloidosis, Hypomagnesemia, Hypophosphatemia
Clinical Presentation	• Asymptomatic hyperuricemia • Acute intermittent → Acute gouty arthritis • Monoarticular • 🕮 Exquisite pain, warm tender swelling—first MTP joint (podagra) most common: – Monoarticular – Other sites: Midfoot, ankles, heels, and knees – Rare: Vertebra(e) mimicking discitis or epidural abscess – Fever, chills, malaise, and cutaneous erythema – May last days to weeks with a mean time of 11 months between attacks • Chronic tophaceous gout: – Tophi form after several years of attacks – Cause structural damage to the articular cartilage and adjacent bone • Polyarticular gout: – Sites of involvement: Olecranon bursae, wrists, hands, renal parenchyma with uric acid nephrolithiasis	• Inflammation in one or more of the large joints • 🕮 Most common—knee • Others: First MTP, wrist, MCP, hips, shoulder, elbow, and crowned dens syndrome • Symmetric • Flexion contracture of the knee is common • Less painful than gout, self-limiting, lasts 2 days to weeks • Fever, chills, and malaise

(*continued*)

TABLE 3–6 Crystal-Induced Synovitis (*continued*)

	GOUT	PSEUDOGOUT
Provocative Factors	• Trauma—influx of synovial fluids urate production • Alcohol—increases uric production • 🕮 Drugs—thiazides, ASA, loop diuretics, niacin • Hereditary	• Hereditary—articular chondrocalcinosis • Idiopathic • Metabolic disease • Trauma • Surgery, illness (MI, CVA)
🕮 Labs	Hyperuricemia	Uric acid normal
Radiologic	Acute gouty arthritis: • Soft tissue swelling around the affected joint • Asymmetric • MTP most frequent joint involved • Others: Fingers, wrists, and elbows Chronic tophaceous • Tophi appear as nodules in lobulated soft tissue masses • Bone erosions develop near the tophi just slightly removed from the periarticular surface, developing overhanging margins • Joint space is preserved • No osteopenia	Chondrocalcinosis: • Punctate fine lines of crystals in the articular hyaline or fibrocartilage tissues • #1—Menisci of the knee: Resulting in calcification of the menisci • Other large joint: Acetabulum labrum, pubic symphysis, articular disc of the wrist, and annulus fibrosis of the disc • Joint effusions
Treatment	Goals → pain relief, prevent attacks, tophi and joint destruction Acute attack: • Colchicine—decreases inflammation by regulating cell proliferation, signal transduction, gene expression, chemotaxis, and PMN degranulation • NSAIDs—Indocin • Corticosteroids • Chronic management (urate-lowering therapy) • Allopurinol and febuxostat—decrease synthesis of uric acid; probenecid may be second line if not tolerated • Probenecid, lesinurad—uricosuric, increase the renal excretion of uric acid • Pegloticase—recombinant uricase dramatically reduces serum uric acid, allowing tophi to dissolve	• NSAIDs • Corticosteroids • Colchicine

ASA, aspirin; CPPD, calcium pyrophosphate dehydrate; CVA, cerebrovascular accident; MCP, metacarpophalangeal; MI, myocardial infarction; MTP, metatarsal phalangeal; NSAID, nonsteroidal anti-inflammatory drug; PMN, polymorphonuclear; WBC, white blood cell.

SERONEGATIVE SPONDYLOARTHROPATHIES

DEFINITION

- Seronegative spondyloarthropathies (SpA) consist of a group of multisystem inflammatory disorders affecting various joints, including the spine, peripheral joints, and periarticular structures.
- Associated with extra-articular manifestations
- Majority are HLA-B27 (+), RF (–), and ANA (–).
- There are four major seronegative SpA **(**Table 3–7**):**
  - Ankylosing spondylitis (AS)
  - Reactive arthritis (formerly Reiter's syndrome)
  - Psoriatic arthritis
  - Arthritis of IBD

🕮 ANKYLOSING SPONDYLITIS

Definition

- Chronic, inflammatory rheumatic disorder of the axial skeleton affecting the sacroiliac (SI) joint and the spine
- Most common symptoms are back pain and significant stiffness, notably in the morning and at night. Symptoms worsen with rest and improve with activity.
- The hallmark is bilateral sacroiliitis.
- Nonradiographic axial spondyloarthritis is chronic inflammatory back pain that has not yet caused erosions of the SI joints on x-rays, but may show up on MRI of the osseous pelvis or have other SpA features like HLA-B27 (+), enthesitis, dactylitis, or uveitis.

HLA-B27 (+) Diseases
- AS
- Reactive arthritis (also known as Reiter's syndrome)
- Psoriatic arthritis—HLA-Cw6
- Enteropathic arthropathy
- Oligoarticular JIA

AS, ankylosing spondylitis; HLA, human leukocyte antigen; JIA, juvenile idiopathic arthritis.

Epidemiology

- Onset → late adolescent and early adulthood
- Males >> females
- More common in Whites
- Genetic marker → HLA-B27 (+) in approximately 90%

Mechanism

- Exact mechanism is unknown.
- Synovitis and inflammation with intimal cell hyperplasia—lymphocyte and plasma cell infiltrate

Ankylosing Spondylitis Versus Rheumatoid Arthritis
Both have synovial inflammation that can lead to destruction of articular cartilage and ankylosis of the joint.

ANKYLOSING SPONDYLITIS	RHEUMATOID ARTHRITIS
More common in males	More common in females
Absence of rheumatoid nodules	Presence of rheumatoid nodules
RF (–) HLA-B27 (+)	RF (+) in 85% of cases HLA-B27 (–)
Marginal syndesmophytes	

HLA, human leukocyte antigen; RF, rheumatoid factor.

Clinical Manifestations

SKELETAL INVOLVEMENT

- Insidious onset of back/gluteal pain:
  - First site of involvement is SI joint. Initially asymmetric but eventually becomes bilateral.
- Persistent symptoms of pain for at least 3 months
- Lumbar morning stiffness that improves with activity and worsens with rest/inactivity
- Reduction of spine flexibility (Positive Shober's test)
- Decreased lumbar lordosis and increased thoracic kyphosis
- 🕮 Cervical ankylosis develops in 75% of patients who have AS for 16 years or longer.
- Lumbar spine or lower cervical is the most common site of fracture.
- Enthesitis: Inflammatory process occurring at the tendon insertion site onto bone
  - Tenderness over the ischial tuberosity, greater trochanter, anterior superior iliac spine (ASIS), and iliac crests
- Hip and shoulder involvement is more common in the juvenile onset, <16 years old.
- Respiratory restriction with limited chest expansion
  - Normal is 7 to 8 cm; if less than that, then there is a risk of restrictive lung disease
  - Once restrictive lung disease pattern ensues:
    - Chest expansion decreases
    - Patient develops diaphragmatic breathing
    - Thoracic spine involvement—costovertebral, costosternal, manubriosternal, and sternoclavicular

Sites of Involvement in AS

1. SI joint
2. Lumbar vertebrae
3. Thoracic vertebrae
4. Cervical vertebrae

AS, ankylosing spondylitis; SI, sacroiliac.

EXTRASKELETAL INVOLVEMENT

- Other complaints include systemic effects—fatigue, weight loss, and low-grade fever.
- Acute iritis (also known as acute anterior uveitis)/iridocyclitis—most common extraskeletal manifestation of AS, and uveitis may present several years before axial symptoms.
- More progressive in Reiter's syndrome:
  - Unilateral, recurrent
  - Pain, photophobia, and blurred vision
- Cardiac:
  - Aortitis leading to fibrosis and aortic root dilatation
  - Conduction defects
- Apical pulmonary fibrosis:
  - May experience dyspnea and cough
- Amyloidosis
- Neurologic:
  - Cauda equina syndrome
  - C1 to C2 subluxation

LAB FINDINGS

- HLA-B27 (+) in 90% of patients
- RF (–) and ANA (–)
- Elevated ESR and CRP
- Anemia—normochromic/normocytic

RADIOGRAPHIC FINDINGS

- SI joint narrowing—symmetric; erosions and sclerosis may lead to ankylosis of the SI joint.
- Pseudo-widening of the joint space:
  - Subchondral bone resorption—blurring of joint line
  - Erosion and sclerosis
  - Calcification leading to ankylosis
- Bamboo spine:
  - Ossification of the annulus fibrosis, resulting in bridging syndesmophytes that completely bridge adjacent vertebral bodies
- Interspinous ligament ossification can give "dagger sign" appearance on x-ray.
- Syndesmophyte formation, spinal ligament ossification, and ankylosis of the facet joints lead to complete spinal fusion.
  - Squaring of lumbar vertebrae's anterior concavity
  - Reactive bone sclerosis
  - Squaring and fusion of the vertebral bodies secondary to ossification of the outer annulus fibrosis at the dorsolumbar and lumbosacral area
- Associated osteopenia/osteoporosis (bone washout)
- Loss of cervical lordosis (straightening of the C-spine)
- Cervical ankylosis and thoracic kyphosis lead to limitations in horizontal gaze, affecting activities such as computer work, cooking, and driving. The chin brow vertical angle (CBVA) is used as a parameter for considering surgery for correction of spinal deformities.
- Hip and shoulder involved to a lesser extent

Schober's Test (Figure 3–4)

- Schober's test is used to detect the limitation of forward flexion and hyperextension of the lumbar spine.
- While standing erect, place a landmark midline at a point 5 cm below the iliac crest line and 10 cm above the spinous processes. On forward flexion, the line should increase by >5 cm to a total of 20 cm or more (from 15 cm).
- Any increase less than 5 cm is considered a restriction.

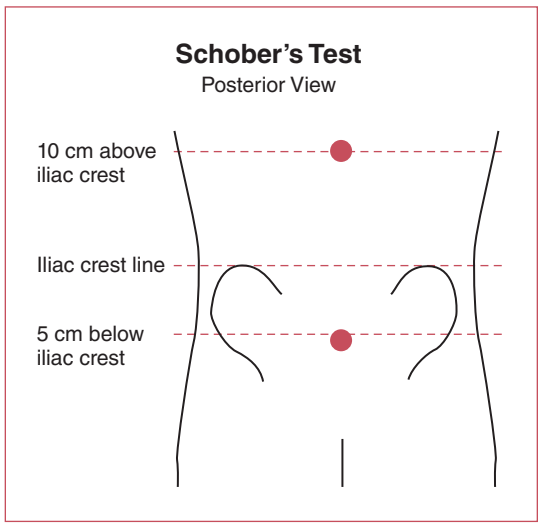

FIGURE 3–4 Schober's test.

Treatment

- Education:
  - Prevent spine flexion contractures
  - Good posture
  - 🕮 Firm mattress, sleep in position to keep spine straight/prevent spine flexion deformity—lie prone
- PT:
  - Spine mobility—extension-based exercises
  - Swimming is ideal.
  - Joint protection

- Pulmonary—maintain chest expansion:
  - Deep breathing exercises
  - Smoking cessation
- Medications:
  - NSAIDs—indomethacin:
    - Control pain and inflammation
    - Allow for PT
  - Corticosteroids—tapering dose, PO, and injections
  - Muscle relaxants
  - DMARDs
    - For patients with purely axial disease, sulfasalazine and methotrexate are not recommended. However, sulfasalazine may be considered in patients who also have peripheral arthritis.
    - 🕮 TNF inhibitors
      - Examples: Etanercept (Enbrel®), infliximab (Remicade®), and adalimumab (Humira®)
      - Mechanism of action: Reduce levels of TNF-alpha (etanercept = soluble receptor; infliximab = chimeric antibody; adalimumab = human monoclonal antibody)
      - Toxicity: Infection (especially TB reactivation), demyelinating disease, induction of autoimmunity, exacerbation of congestive heart failure, possible malignancy (skin CA)
      - Selective IL-17A inhibitors
        - Examples: Secukinumab (Cosentyx®) and ixekizumab (Taltz®)
        - Mechanism of action: Block IL-17A receptor (human monoclonal antibodies)
        - Toxicity: Nasopharyngitis, infections, induction or worsening of IBD (contraindicated in known IBD)
  - Topical corticosteroid drops—uveitis

REACTIVE ARTHRITIS (FORMERLY REITER'S SYNDROME)

🕮 Triad of Reactive Arthritis

1. Conjunctivitis
2. Arthritis
3. Nongonococcal urethritis

("Can't see, can't pee, can't climb a tree.")

Epidemiology

- Males >> females
- Typically follows GI or genitourinary (GU) infection
- Organisms (two main groups):
  - Sexually transmitted diseases (STDs): *Chlamydia*
  - GI infection: *Campylobacter*, *Yersinia*, *Shigella*, *Salmonella*
  - Also associated with HIV
- More common in Caucasian population
- 🕮 Approximately 3% to 10% of patients with reactive arthritis progress to AS.

🕮 Clinical Manifestations

ARTHRITIS

- Arthritis appears 2 to 4 weeks after initiating infectious event—GU or GI.
- 🕮 Asymmetric
- Oligoarticular—average of four or fewer joints:
  - LE involvement >> UE
  - More common in the knees, ankles, and small joints of the feet

  - 🕮 May be confused with plantar fasciitis
  - Rare hip involvement
  - UE → wrist, elbows, and small joints of the hand
- Sausage digits (dactylitis):
  - Swollen, tender digits with a dusk-like blue discoloration
  - Pain on ROM
- Enthesopathies—Achilles tendon:
  - Swelling at the insertion of tendons, ligaments, and fascia attachments
- Low back pain—sacroiliitis

OCULAR
- Conjunctivitis, iritis, uveitis, episcleritis, corneal ulceration

GENITOURINARY
- Urethritis, meatal erythema, edema
- Balanitis circinata—small painless ulcers on the glans penis or urethritis

SKIN AND NAILS
- Keratoderma blennorrhagica—hypertrophic skin lesions on palms and soles of feet
- Reiter's nails—thickened and opacified, crumbling, and nonpitting

CARDIAC
- Conduction defects

GENERAL
- Weight loss, fever
- Amyloidosis

Lab Findings
- Synovial fluid: Inflammatory changes
- Positive evidence of GI or GU infection
- Increased ESR
- RF (–) and ANA (–)
- Anemia—normochromic/normocytic
- HLA-B27 (+)

Reactive Arthritis: Inflammatory Synovial Fluid
- Turbid
- Poor viscosity
- WBC 5,000–50,000 PMNs
- Increased protein, normal glucose

PMN, polymorphonuclear; WBC, white blood cell.

🕮 Radiographic Findings
- "Lover's heel"— erosion and periosteal changes at the insertion of the plantar fascia and Achilles tendons
- Ischial tuberosities and greater trochanter
- Asymmetric SI joint involvement
- Syndesmophytes
- Pencil-in-cup deformities of the hands and feet—more common in psoriatic arthritis

Treatment
- NSAIDs such as indomethacin
- Antibiotics: Typically tetracycline or erythromycin-based
- Corticosteroids
- DMARDs

PSORIATIC ARTHRITIS

Prevalence

- Approximately 5% to 7% of persons with psoriasis will develop some form of inflammatory joint disease.
- Affects 0.1% of the population
- Equal male:female ratio
- Age of onset ranges between 30 and 55 years.
- More common in Whites
- Associated with HIV

Psoriatic Arthritis and HIV

- Foot and ankle involvement is most common and severe.
- Treatment—same as psoriatic:
  - First-line NSAIDs
  - No oral corticosteroids
  - No methotrexate

NSAID, nonsteroidal anti-inflammatory drug.

Pathogenesis

- Unknown
- Genetic—HLA-B27 (+)
- Environmental—infectious, trauma
- Immunologic

Clinical Manifestations

SKIN AND NAILS

- Psoriatic skin lesions—erythematous, silvery scales
- Auspitz's sign—gentle scraping of the lesions results in pinpoint bleeding
- Located over the extensor surfaces
- Nail pitting

ARTHRITIS

- Stiffness of the spine lasting approximately 30 minutes
- Asymmetric monoarticular or oligoarticular involvement:
  - Large joints → knee
  - DIP involvement
    - Arthritis mutilans—osteolysis of the phalanges and metacarpals of the hand resulting in "telescoping of the finger"
- Enthesopathy: Inflammation of the enthesis (insertion of ligament, tendon, joint capsule, and bone)
- Spondylitis, sacroiliitis

OTHER SYSTEMIC INVOLVEMENT

- Conjunctivitis—one-third
- Aortic insufficiency

Lab Findings

- Nonspecific—increased incidence in patients with HLA-B27 (+)

Radiographic Findings

- "Pencil-in-cup" appearance of the DIP
- Asymmetric sacroiliitis → fusion
- "Fluffy periostitis"—hands, feet, spine, and SI joint
- Syndesmophytes—see Radiographic Findings under Ankylosing Spondylitis section
- Bone erosion

🕮 Treatment

- ROM to all joints
- Do not abuse an inflamed joint → exacerbation
- Medications—similar to RA, psoralen plus ultraviolet A (PUVA; long wave ultraviolet Å light)
  - Steroids—oral steroids not proven, injection may help
  - Biologicals: Anti-TNF antibodies (adalimumab, infliximab) work best.

ENTEROPATHIC ARTHROPATHY

Definition

- Inflammatory SpA associated with IBD (Crohn's disease or ulcerative colitis) and reactive arthritis (bacterial etiology)

Epidemiology

- Male >> female
- Peripheral arthritis occurs in approximately 10% to 20% of patients with Crohn's disease and ulcerative colitis.

Clinical Manifestations

- Asymmetric joint involvement
- Synovitis affecting the peripheral joints
- Monoarticular or polyarticular
- 🕮 Large joints—knees, ankles, and feet
- Two types of arthropathies can occur:
  - Enteropathic arthritis
  - AS
- Sacroiliitis
- Peripheral arthritis will subside with remission of bowel disease.

TABLE 3–7 Seronegative Spondyloarthropathy Fact Sheet

The following are all seronegative spondyloarthropathies:
1. Ankylosing spondylitis
2. Reactive arthritis (formerly Reiter's syndrome)
3. Psoriatic arthritis
4. Arthritis of inflammatory bowel disease

ALL HAVE THE FOLLOWING CHARACTERISTICS:	ANKYLOSING SPONDYLITIS	REACTIVE ARTHRITIS[a]	PSORIATIC ARTHRITIS	ENTEROPATHIC ARTHROPATHIES
1. RF (–)	✓	✓	✓	✓
2. Increased incidence in patients with (+) HLA-B27	✓	✓	✓	✓
3. Mucocutaneous lesions	✓	✓	✓	✓
4. Frequent inflammation of the enthesis	✓	✓	✓	✓
5. Spondylitis with SI joint involvement	✓	✓	✓	✓

[a]Reactive arthritis formerly known as Reiter's syndrome.
HLA, human leukocyte antigen; RF, rheumatoid factor; SI, sacroiliac.

Extra-Articular Manifestations

- Erythema nodosum—Crohn's
- Pyoderma gangrenosa—ulcerative colitis
- Painful deep oral ulcers
- Uveitis
- Fever and weight loss during bowel episodes

Lab Test

- Anemia
- Elevated ESR and CRP
- RF (–), ANA (–)
- (+) Antineutrophil cytoplasmic antibodies (ANCAs) approximately 60% (antimyeloperoxidase)
- Increase incidence of HLA-B27 (+)

OTHER RHEUMATOID DISEASES

SYSTEMIC LUPUS ERYTHEMATOSUS

- Multisystemic, autoimmune disease that affects every organ in the body
- Systemic vascular inflammation caused by an autoimmune response of unknown etiology
- Female >> male
- The ACR and EULAR Criteria for SLE (updated 2019) require an ANA of at least 1:80 and a total score of at least 10 across the domains (see below); the highest weighted criterion is counted from each domain.

ACR AND EULAR CRITERIA FOR SLE
ENTRY CRITERIA: ANA >1:80

CLINICAL DOMAINS AND CRITERIA	Wt
Constitutional	
Fever	2
Hematologic	
Leukopenia	3
Thrombocytopenia	4
Autoimmune hemolysis	4
Neuropsychiatric	
Delirium	2
Psychosis	3
Seizure	5
Mucocutaneous	
Nonscarring alopecia	2
Oral ulcers	2
Subacute cutaneous or discoid lupus	4
Acute cutaneous lupus (malar rash)	6

IMMUNOLOGIC DOMAINS AND CRITERIA	Wt
Antiphospholipid antibodies	
Anti-cardiolipin antibodies OR Anti-B2GP 1 antibodies OR Lupus anticoagulant	2
Complement proteins	
Low C3 OR low C4	3
Low C3 AND low C4	4
SLE-specific antibodies	
Anti-dsDNA antibody OR Anti-Smith antibody	6

(*continued*)

CLINICAL DOMAINS AND CRITERIA	Wt
Serosal	
Pleural or pericardial effusion	5
Acute pericarditis	6
Musculoskeletal	
Joint involvement	6
Renal involvement	
Proteinuria >0.5 g/24 hr	4
Renal biopsy Class II or V lupus nephritis	8
Renal biopsy Class III or IV lupus nephritis	10

ACR, American College of Rheumatology; ANA, antinuclear antibody; EULAR, European League Against Rheumatism; SLE, systemic lupus erythematosus; Wt, weight.
Source: From Aringer M, Costenbader K, Daikh D, et al. 2019 European League Against Rheumatism/American College of Rheumatology classification criteria for systemic lupus erythematosus. *Arthritis Rheumatol*. 2019;71(9):1400–1412. doi:10.1002/art.40930.

Mnemonic to remember criteria: DOPAMINE RASH: Discoid rash, **O**ral ulcers, **P**hotosensitivity, **A**rthritis, **M**alar (butterfly) rash, **I**mmunologic disorder, **NE**urologic disorder, **R**enal disorder, **A**bnormal ANA titer, **S**erositis, **H**ematologic disorder

Clinical Features

- Fatigue, fever, weight loss, and GI complaints
- Alopecia
- Vasculitis
- Arthritis:
  - Small joints of the hands, wrist, and knees
  - Symmetric
  - Migratory, chronic, and nonerosive
  - Soft tissue swelling
  - Subcutaneous nodules
  - Synovial analysis
  - Jaccoud's arthritis
- Arthralgias
- Muscle pain and weakness

Jaccoud's Arthritis
- Nonerosive deforming arthritis
- Ulnar deviation of the fingers and subluxations that are reversible early and do not typically cause functional impairment
- May become fixed

Labs

- Depressed complement levels—C3 and C4
- Anti-ds DNA: Specific for SLE
- Anti-Sm: Specific for SLE
- ANA with homogenous, speckled, or nucleolar pattern
- Cytopenias
- Proteinuria

Treatment

- NSAIDs, corticosteroids, antimalarials, methotrexate, cyclophosphamide, azathioprine, cyclosporine A, dapsone, belimumab, and possibly rituximab

SCLERODERMA (SYSTEMIC SCLEROSIS)

- Progressive chronic multisystem disease characterized by three hallmarks: Small vessel vasculopathy, production of autoantibodies, and fibroblast dysfunction leading to increased deposition of extracellular matrix
- Results in skin thickening with various involvement of internal organs
- Classification criteria updated in 2013 by ACR/EULAR (van den Hoogen et al., 2013):
  - Presence of skin thickening of the fingers extending proximal to the MCP joints. Other symptoms that may be present and carry varying weights include (see van den Hoogen et al. for further details) skin thickening of the fingers, fingertip lesions, telangiectasia, abnormal nailfold capillaries, pulmonary arterial hypertension, Raynaud's phenomenon, and systemic sclerosis (SSc)-related autoantibodies.
- Fibrosis-like changes in the skin and epithelial tissues of affected organs
- Subsets:
  - Diffuse cutaneous scleroderma:
    - Skin thickening above and below the knees and elbows and also involving the face and trunk
    - Heart, lung, GI, and kidney
    - ANA (+)
    - Anticentromere antibody (–)
    - Rapid onset after Raynaud's phenomenon
    - Variable course—poor prognosis
  - Limited cutaneous scleroderma—CREST syndrome:
    - Skin thickening limited to the face and below the elbows (forearms, hands, and fingers) and knees (lower legs and feet)
    - Progression after Raynaud's phenomenon
    - Anticentromere antibody (+)
    - Good prognosis
  - Overlap syndromes:
    - Combinations of CTD
  - Undefined CTD:
    - No clinical or laboratory findings
  - Localized scleroderma:
    - Morphea and linear scleroderma

Eosinophilic Fasciitis (may look similar to scleroderma but is not related)
- Precipitated by strenuous exercise
- Exercise should be done in a noninflammatory state
- Pain and swelling
- Treatment—steroids

Crest Syndrome
- **C**alcinosis
- **R**aynaud's phenomenon
- **E**sophageal dysmotility
- **S**clerodactyly
- **T**elangiectasia

Clinical Features

- Skin thickening—face, trunk, and neck
- Symmetric arthritis with involvement of the fingers, hands, arm, and legs
- Initial symptoms—Raynaud's phenomenon with fatigue and musculoskeletal complaints (such as generalized pain and stiffness)
- In diffuse scleroderma, erosive arthritis of the fingers can occur with distal bone resorption, osteolysis, and calcinosis. Limited scleroderma may present solely as grip weakness and limited finger mobility (Wigley & Boin, 2017).

RAYNAUD'S PHENOMENON

- Vasospasm of the muscular digital arteries that can lead to ischemia and ulceration of the fingertips
- Triggered by cold and emotional stresses

- Reversal of episode occurs after stimulus has ended—and digits rewarmed.
- Present in 90% of patients with scleroderma
- Treatment:
  - Education against triggers—cold, smoking
  - Rewarming
  - Calcium channel blockers—nifedipine, amlodipine
  - Topical nitroglycerin paste
  - Phosphodiesterase 5 inhibitors—sildenafil, tadalafil
  - IV prostacyclin analogues—iloprost
  - Digital sympathectomy
  - Electromyography (EMG) and biofeedback—self-regulation

Causes of Raynaud's Phenomenon

- Collagen vascular disease—PSS, SLE, RA, and dermatomyositis/polymyositis
- Arterial occlusive disease
- Pulmonary hypertension
- Neurologic—SCI, CVA
- Blood dyscrasia
- Trauma
- Drugs—ergots, beta-blockers, cisplatin (Braunwald et al., 2001)

CVA, cerebrovascular accident; PSS, progressive systemic sclerosis; RA, rheumatoid arthritis; SCI, silent cerebral infarction; SLE, systemic lupus erythematosus.

Treatment

- ROM exercises twice daily
- Strengthening exercises
- Increase skin elasticity

POLYMYOSITIS/DERMATOMYOSITIS

- Inflammatory myopathies involving striated muscle and clinically presents with profound symmetrical weakness of the proximal muscles
  - Shoulder and pelvic girdle
  - Anterior neck flexors
  - 🕮 Pharyngeal involvement → dysphagia results

Five Types

- **Type I**—primary idiopathic polymyositis; insidious onset
  - Weakness starts at the pelvic girdle → shoulder girdle → neck
  - Dysphagia/dysphonia
  - Remission and exacerbation common
  - Moderate-severe arthritis
  - Atrophic skin over knuckles
- **Type II**—primary idiopathic dermatomyositis; acute onset
  - Proximal muscle weakness, tenderness
  - Heliotrope rash with periorbital edema
  - Arthritis
  - Raynaud's phenomenon
  - ILD
  - Malaise, fever, and weight loss
- 🕮 **Type III**—dermatomyositis or polymyositis; 5% to 8% associated with malignancy
  - Male >40 years old
  - Poor prognosis
  - Anti-TIF-1-gamma and anti-NXP-2 are specific antibodies associated with greatly increased risk of malignancy.
- **Type IV**—childhood dermatomyositis or polymyositis
  - Rapid progressive weakness
  - Respiratory weakness
  - 🕮 Severe joint contractures—more disabling in a child
- **Type V**—polymyositis or dermatomyositis; associated with collagen vascular disease

🕮 Clinical Features of Polymyositis/Dermatomyositis—Modified American College of Rheumatology Criteria

- Symmetric proximal muscle weakness:
  - Hips involved first, then shoulders
  - (+/–) Respiratory muscle involvement
  - Dysphagia
- Muscle biopsy:
  - Perifascicular atrophy
  - Evidence of necrosis of type I and II fibers
  - Variation in fiber size
  - Large nuclei
- Elevation of muscle enzymes:
  - Elevated creatinine phosphokinase, aldolase levels; elevated transaminases and lactate dehydrogenase (LDH)
- ACR scoring criteria: Minimum score of 5.5 required for diagnosis (see table below)

ACR Scoring Criteria for Polymyositis/Dermatomyositis

CLINICAL SIGN	SCORE
Age of onset of first symptom: 18–40 years old	1.3
Age of onset of first symptom: >40 years old	2.1
Objective symmetric muscle weakness of proximal upper extremities	0.7
Objective symmetric muscle weakness of proximal lower extremities	0.8
Neck flexors relatively weaker than neck extensors	1.9
Proximal leg muscles relatively weaker than distal leg muscles	0.9
Heliotrope rash	3.1
Gottron's papules	2.1
Gottron's sign	3.3
Dysphagia/esophageal dysmotility	0.7
Anti-Jo-1 (+)	3.9
Elevated CPK or LDH or AST/ALT	1.3
Muscle biopsy with endomysial infiltration of mononuclear cells surrounding but not invading myofibers	1.7
Muscle biopsy with perimysial and/or perivascular infiltration of mononuclear cells	1.2
Muscle biopsy with perifascicular atrophy	1.9
Muscle biopsy with rimmed vacuoles	3.1

ACR, American College of Rheumatology; ALT, alanine aminotransferase; AST, aspartate aminotransferase; CPK, creatine phosphokinase; LDH, lactate dehydrogenase.

Source: From Lundberg IE, Tjarnlund A, Bottai M, et al. 2017 European League Against Rheumatism/American College of Rheumatology Classification Criteria for Adult and Juvenile Idiopathic Inflammatory Myopathies and Their Major Subgroups. *Arthritis Rheumatol*. 2017;69(12):2271–2282. doi:10.1002/art.40320 © 2017, American College of Rheumatology.

- 🕮 EMG:
  - Myopathic changes:
    - Small-amplitude, short-duration polyphasic motor units
    - Early recruitment pattern
  - In addition:
    - Positive sharp waves, fibrillation potentials
    - Complex repetitive discharges (CRD)
- Dermatologic features—dermatomyositis:
  - Lilac heliotrope rash with periorbital edema
  - Gottron's papules—scaly dermatitis over the dorsum of the hand—MCP, PIP
  - Other photosensitive areas that develop erythematous macular rash: Sleeve sign (upper arm), holster sign (lateral hip), shawl sign (posterior neck/upper back), V sign (anterior neck and chest)

Poor Prognostic Factors

- Old age
- Malignancy
- Cardiac involvement
- Delayed initiation of corticosteroid therapy
- Respiratory muscle weakness—aspiration pneumonia
- Joint contractures
- ILD

Treatment

- Corticosteroids: Generally 1 mg/kg/day prednisone for 4 to 6 weeks, then taper
- Second line—azathioprine or methotrexate
- IV immunoglobulin in severe, refractory cases (myositis, ILD, and dysphagia)
- ROM, isometric exercises—defer strengthening exercises until inflammation controlled
- Follow—serum enzymes and manual muscle strength testing

🕮 Juvenile Dermatomyositis

- Seen more commonly than polymyositis in children
- Associated with generalized vasculitis (unlike adult form)
- Slight female preponderance
- Heliotrope rash is a predominant feature.
- Presence of clumsiness is often unrecognized.
- Clinically—transient arthritis, elevated rash
- 80% to 90% respond well to corticosteroids.
- No association with malignancy in children

MIXED CONNECTIVE TISSUE DISORDERS

- Mixed connective tissue disorders (MCTDs) refer to disorders with characteristics of several other diseases, in particular:
  - SLE
  - Scleroderma (SSc)
  - Polymyositis
- Overlapping symptoms include:
  - Raynaud's phenomenon
  - Synovitis in the joints of the hand
  - Arthritis
  - Puffy hands

- Myopathy
- Esophageal dysmotility
- Acrosclerosis
- Pulmonary hypertension
- Abnormal antibodies

KEY POINTS OF ARTHRITIDES

- The following table indicates in what circumstances ANA and RF are positive or negative.

🕮 ARTHRITIDES: ANTINUCLEAR ANTIBODY AND RHEUMATOID FACTOR STATUS

SYNDROME	ANA	RF
MCTD	+	+
RA	+	+
SLE	+	–
Scleroderma (PSS)	+	–
Polymyositis	+	–
Sjögren's syndrome	+	+

ANA, antinuclear antibody; MCTD, mixed connective tissue disorder; PSS, progressive systemic sclerosis; RA, rheumatoid arthritis; RF, rheumatoid factor; SLE, systemic lupus erythematosus.

🕮 HLA-B27 (+) SYNDROMES

- AS
- Reactive arthritis (formerly Reiter's syndrome)
- Psoriatic arthritis
- Enteropathic arthropathy
- Oligoarticular JIA

🕮 STIFF PERSON SYNDROME

- Postulated to be an autoimmune-mediated disorder against gamma-aminobutyric acid (GABA) synthesis resulting in a loss of inhibition at the brainstem and spinal cord
- Characterized by muscle stiffness, rigidity, and painful muscle spasms
  - Trunk musculature typically affected first. Lower extremities more commonly affected than upper extremities.
  - Spasms can be triggered by environmental stimulation such as loud noises, physical touch, changes in temperature, and stressful events.
- Clinical manifestations:
  - Gait difficulty secondary to LE stiffness or spasms
  - Axial muscle rigidity and spasms can cause lumbar lordosis, scapular muscle spasms, mid-thoracic back pain, and abdominal pain. This mimics AS in posture and presentation, although the etiology is muscular, not arthritis.
- Epidemiology:
  - Affects one out of every million people
  - Affects females twice as likely compared with males
  - Symptoms most commonly develop between the ages of 30 and 50, but pediatric cases have been documented as well.

- Associated with other autoimmune conditions: Type 1 diabetes, autoimmune thyroid disease, vitiligo, pernicious anemia, and celiac disease
- Lab findings:
  - Antibodies against glutamic acid decarboxylase (GAD), which is a protein that inhibits GABA, can be seen in both serum and CSF.
- EMG studies will show impaired inhibition of H-reflexes and continuous motor unit activity, which is a characteristic finding.
- Treatment is focused on improving symptomology of muscle spasms and muscular pain.
  - Benzodiazepines: GABA receptor agonists. Diazepam is first-line treatment.
  - Oral baclofen has also been reported in the literature to help with symptomatology. However, some patients have required intrathecal baclofen due to significant side effects and lack of central nervous system penetration of oral baclofen.
  - Botulinum toxin injections
  - Neuropathic pain medications: Gabapentin and pregabalin can also be used.
  - Immunotherapy: Intravenous immunoglobin and rituximab have been used with good effect.

VASCULITIDES

LARGE VESSEL VASCULITIDES

Takayasu Arteritis

- Affects the large arteries—aorta, arch, and main branches
- Asian females, 40 years old
- Signs/symptoms:
  - Erythema nodosum on the legs
  - Pulselessness, arm claudication

Temporal Arteritis

- Also known as giant cell arteritis (GCA)
- Involves large arteries
- More common in females >50 years old
- Symptoms:
  - Tenderness of the scalp and in the muscle of mastication
  - Headaches
  - Abrupt visual loss in 15% of patients
  - Associated with polymyalgia rheumatica (PMR; see the following section)
- Diagnosis: Elevated ESR, temporal artery biopsy
- 🕮 Treatment: High-dose steroids ASAP imperative to prevent permanent vision loss, ASA (325 mg daily—improves prognosis)

🕮 Polymyalgia Rheumatica

- In view of clinical similarities between PMR patients with and without signs of vasculitis in a temporal artery biopsy, many authors believe that PMR is an expression of temporal arteritis.
- Up to 16% of PMR patients develop temporal arteritis, and 50% of temporal arteritis patients have PMR symptoms.
- Symptoms include:
  - Fever, weight loss, malaise
  - Morning stiffness—muscle tenderness
  - Hallmark—difficulty abducting shoulders above 90 degrees
  - Affects proximal muscles—neck, pelvic

- Abrupt myalgias/arthralgia
- Diagnosis: ESR >50
- Treatment: Low-dose prednisone (maximum 15–20 mg daily) with resolution of symptoms in most patients within 2 to 3 days and in all patients by 14 days; reduce to 10 mg daily by weeks 4 to 8; then taper by 1 mg/month until off of steroids
- Relapse (during steroid taper) and recurrence (when off of steroids) are common in the natural history of PMR.

MEDIUM VESSEL VASCULITIDES

Polyarteritis Nodosa

- Systemic necrotizing vasculitis involving small-to-medium-sized arteries
- 2:1 male:female ratio
- Glomerulonephritis—#1 cause of death
- Lungs spared
- Skin—palpable purpura
- GI and GU—bowel ischemia (postprandial abdominal pain, nausea/vomiting, bleeding, intestinal or renal artery stenosis or aneurysm)
- Mononeuritis multiplex, arthritis

Polyarteritis Nodosa Also Seen In:
- RA
- SLE
- Sjögren's syndrome

RA, rheumatoid arthritis; SLE, systemic lupus erythematosus.

ANTINEUTROPHIL CYTOPLASMIC ANTIBODY-ASSOCIATED VASCULITIDES

Granulomatous Vasculitis (Formerly Wegener's Granulomatosis)

- Small-to-medium-sized artery involvement
- More common in middle-aged males
- Necrotizing granulomatous vasculitis involving:
  - Upper/lower respiratory tract
  - Pauci-immune glomerulonephritis
- "Saddle-nose" deformity
- Pulmonary, tracheal, ocular, and cutaneous manifestation

Microscopic Polyarteritis

- Small-to-medium-sized arteries involved
- Few or no immune deposits seen
- Renal and pulmonary involvement

Churg–Strauss Syndrome (Eosinophilic Granulomatosis with Polyangiitis)

- Eosinophil-rich and granulomatous inflammation
- Small-to-medium-sized arteries involved
- Respiratory tract involvement predominates:
  - Associated with asthma, eosinophilia
- Neuropathy common

OTHER VASCULITIDES

Behçet's Syndrome

- Small vessels involved
- Oral and genital skin ulcers
- 20% of patients experience venous thrombosis
- Neurologic manifestations can include ischemic stroke, cranial and optic neuropathy, meningoencephalitis, venous sinus thrombosis, intracranial hypertension, and uveitis.

Goodpasture's Syndrome

- Pulmonary and kidney involvement
- Caused by antibodies to glomerular basement membrane

SJÖGREN'S SYNDROME

Sjögren's syndrome is an autoimmune-mediated disorder of the exocrine glands.

🕮 CLINICAL PRESENTATION (SICCA SYMPTOMS)

- Dry eyes
- Dry mouth
- Skin lesions
- Parotid involvement
- Primary Sjögren's syndrome occurs in people with no other rheumatologic disorders.
- Secondary Sjögren's syndrome occurs in patients with other rheumatologic disorders, most commonly RA and SLE.

LABS

Classification:

- Primary—dry eyes and mouth with ANA (+), RF (+)
- Secondary—sicca symptoms
  - Sjögren's syndrome plus evidence of SLE, RA, progressive systemic sclerosis (PSS), or polymyositis

EXTRAGLANDULAR MANIFESTATIONS

- Arthralgias
- Raynaud's phenomenon
- Neuropathy
- ILD

INFECTIOUS ARTHRITIDES

SEPTIC ARTHRITIDES

- Clinical picture of septic arthritis:
  - Rapid onset of moderate to severe joint pain, erythema, and decreased ROM
  - Monoarticular, leukocytosis
  - Knee is the most common joint
  - Fevers/chills, sepsis
- Risk factors:
  - Age
  - Prosthetic joints/foreign body
  - Comorbidities such as anemia, chronic diseases, and hemophilia
- 🕮 Organisms:
  - *Neisseria gonorrhoeae* → most common in adults
  - *Staphylococcus aureus* → most common in children

Septic Arthritis in Children

CAUSES

- Otitis, infected IV lines
- Neonates and >2 years old: *S. aureus* and group B strep
- 6 months to 2 years old: *Haemophilus influenzae*

PRESENTATION

- Large joints, monoarticular
- Polyarticular infections

Septic Arthritis in Adults/Older Adults

CAUSES

- In adults → ≤60 years of age, main cause is from an STD
- In adults >60 years of age—source is commonly from another focus
- *N. gonorrhoeae*—most common form of acute bacterial arthritis

Rheumatoid Arthritis

- *S. aureus* is the most common organism causing septic arthritis in RA.

🕮 MOST COMMON ORGANISMS IN SEPTIC ARTHRITIS

NEONATES	6 MONTHS TO 2 YEARS OLD	CHILDREN >2 YEARS OLD	ADULTS	RA
S. aureus Group B strep	*Haemophilus influenzae*	*S. aureus* Group B strep	*Neisseria gonorrhoeae*	*S. aureus*

RA, rheumatoid arthritis; *S. aureus*, *Staphylococcus aureus*.

Diagnostic Approach

- Synovial fluid analysis—most important test (Table 3–8)
- Lab work: Elevated white blood cells (WBC), ESR, and CRP
- Radiographic findings:
  - Early: Soft tissue swelling
  - Later: Joint space narrowing, erosions, gas formation (*Escherichia coli, Clostridium perfringens*)
- Bone scans

TABLE 3–8 Joint Fluid Analysis

PROPERTY	NORMAL	NONINFLAMMATORY	INFLAMMATORY	SEPTIC
Viscosity	High	High	Low	Variable
Color	Colorless	Straw	Yellow	Turbid-yellow
Clarity	Translucent	Translucent	Translucent/opaque	Opaque
White blood cell	<200	<5,000	1,000–75,000	>100,000
PMN%	<25	<25	>50	>85
Culture	Negative	Negative	Negative	Positive
Mucin clot	Good	Good	Fair	Poor
Glucose	Equal to blood	Equal to blood	<50 mg/dL lower than blood glucose	>50 mg/dL lower than blood glucose

PMN, polymorphonuclear.

Treatment

- IV antibiotics
- May require serial needle aspirations and/or arthroscopic lavage

Other Causes of Septic Arthritis

- **Viral infections**—rubella, infectious hepatitis
- **Fungi**—seen in immunocompromised adults
- **Mycobacterium TB:**
  - TB spondylitis (Pott's disease):
    - Most commonly seen in lower thoracic/upper lumbar regions
    - Anterior vertebral body preferentially affected, resulting in kyphotic deformities
  - 🕮 TB arthritis—hips and knees:
    - Monoarticular
    - Radiologic findings—Phemister's triad
    - Juxta-articular osteoporosis
    - Marginal erosions
    - Joint space narrowing
- **Lyme disease:**
  - Tick-borne infection from *Borrelia burgdorferi*
  - Classic presentation:
    - Erythema migrans ("bull's eye" rash)
    - Cardiac, neurologic, articular manifestations
    - Pattern of onset: Bite → rash → systemic disease → neurologic involvement
  - 🕮 Intermittent migratory episodes of polyarthritis
  - Commonly affects the knee
  - Synovial fluid—inflammatory. Synovial fluid polymerase chain reaction (PCR) is highly sensitive (>75%) prior to antibiotic treatment and may also indicate if Lyme arthritis persists after antibiotic treatment.
  - Diagnosis—enzyme-linked immunosorbent assay (ELISA), Western blot analysis
  - Management—first-line antibiotics:
    - Adults: Doxycycline
    - Children: Amoxicillin

DEPOSITION/STORAGE DISEASE-RELATED ARTHRITIDES

HEMOCHROMATOSIS

- Organ damage and tissue dysfunction secondary to excessive iron stores and the deposition of hemosiderin
- Organs → hepatic cirrhosis, cardiomyopathy, diabetes mellitus, pituitary dysfunction
- Skin hyperpigmentation
- Chronic progressive arthritis:
  - Occurs commonly in the second and third MCP, PIP joints. It may also affect the hip joints.
- Males approximately 40 to 50 years old
- Treatment: Phlebotomy, NSAIDs

ALKAPTONURIA (OCHRONOSIS)

- Autosomal recessive
- Deficiency in the enzyme homogentisic acid oxidase leads to its increase.
- Alkalinization and oxidation cause darkening of tissue parts, termed ochronosis.

  - Bluish discoloration of the urine, cartilage, skin, and sclera secondary to the accumulation of homogentisic acid
- Progressive degenerative arthropathy:
  - Onset in the fourth decade
  - Spinal column involvement
  - Arthritis of the large joints, chondrocalcinosis, effusions, and osteochondral bodies

WILSON'S DISEASE

- Autosomal recessive
- Deposition of copper leads to destruction.
  - Liver leading to cirrhosis
  - Brain
  - Kidneys
  - Ocular—Kayser–Fleischer rings
- OA—wrists, MCP, knees, and spine
- Osteoporosis
- Treatment: Copper chelation with penicillamine, dietary restriction

GAUCHER'S DISEASE

- Autosomal recessive—common in Ashkenazi Jews
- Glucocerebroside accumulates in the reticuloendothelial cells of the spleen, liver, and bone marrow.
- Monoarticular hip and knee degeneration

OTHER SYSTEMIC DISEASES WITH ARTHRITIS

SARCOIDOSIS

- Systemic chronic granulomatous disease—can affect any organ system
- Pathogenesis: Disseminated noncaseating granulomas
- 8× more common in Blacks
- Females > males
- Clinical features:
  - Pulmonary (nodules, bronchiectasis, and fibrosis)
  - Hilar adenopathy
  - Fever, weight loss, fatigue
  - Arthritis: Polyarthritis, four to six joints:
    - Knees, PIP, MCP, and wrists
  - Skin—Lofgren's syndrome (arthritis, hilar adenopathy, and erythema nodosum)

AMYLOIDOSIS

- Deposition of amyloid in the kidneys, liver, and spleen
- Homogeneous eosinophilic material seen with Congo red dye
- Clinical features:
  - Renal disease is primary clinical feature.
  - Cardiomyopathy
  - Median neuropathy
  - Pseudoarthritis—periarticular joint inflammation
  - Effusions: Arthrocentesis—"shoulder-pad" sign

HEMOPHILIC ARTHROPATHY

- Hemophilia is a blood coagulation disorder caused by factor VIII deficiency (classic hemophilia A) or factor IX deficiency (Christmas disease, hemophilia B).
- X-linked recessive disorder → predominantly in males
- Also associated with HIV secondary to transfusions of factor and blood
- Bleeding into bones and soft tissue causes hemarthrosis, necrosis, and compartment syndrome:
  - Elbows, knees, and wrists commonly involved
  - 🕮 Arthritis caused by the remaining blood in the joint depositing hemosiderin into the synovial lining → synovial proliferation and pannus formation
- Radiographs:
  - Joint space narrowing
  - Subchondral sclerosis
  - Cyst formation
- 🕮 Treatment:
  - Conservative care (immobilization, rest, and ice), factor VIII replacement, rehabilitation
  - 🕮 Joint aspiration as a last resort. Blood in the joint acts as a tamponade to prevent further bleeding.

SICKLE CELL DISEASE

- Biconcave red blood cell (RBC) changes to an elongated crescent sickle shape due to abnormal hemoglobin S protein, causing obstruction of the microvasculature.
- Autosomal recessive inheritance
- Musculoskeletal complications:
  - Painful crisis—most common event:
    - Abdomen, chest, and back
    - Pain in the large joints from juxta-articular bone infarcts with synovial ischemia
  - Dactylitis: "Hand-foot" syndrome:
    - Painful, nonpitting swelling of the hands and feet
  - Osteonecrosis (avascular necrosis):
    - Local hypoxia with occlusion to the venous system by the sickled cells
    - One-third of femoral heads and one-fourth of humeral heads go on to develop osteonecrosis.
  - Osteomyelitis—most commonly caused by *Salmonella*

CHARÇOT JOINT (NEUROPATHIC ARTHROPATHY)

DEFINITION

- A Charçot joint is a chronic, progressively degenerative arthropathy secondary to a sensory neuropathy (loss of proprioception and pain sensation) that leads to joint instability and destruction.

🕮 CAUSES → "STD" → "SKA" (SHOULDER, KNEE, AND ANKLE)

- **S**yringomyelia → **S**houlder
- **T**abes dorsalis → Syphilis → **K**nee
- **D**iabetic neuropathy → #1 cause → **A**nkle

🕮 Charçot Joint Versus Osteoarthritis

They may resemble each other early on.

- Both have:
  - Soft tissue swelling
  - Osteophytes
  - Joint effusion
- Charçot joints have:
  - Bony fragments
  - Subluxation
  - Periarticular debris

CLINICAL FEATURES

- Early findings: Painless swelling, effusion, and joint destruction
- Late findings: Crepitation, destruction of cartilage and bones, intra-articular loose bodies
- Subtle fractures

RADIOGRAPHIC FINDINGS

- Joint destruction
- Hypertrophic osteophytes
- Loose bodies caused by microfractures
- Disorganization of the joint—subluxation and dislocation

TREATMENT

- Immobilization/bracing
- Restriction of weight-bearing

ATRAUMATIC HIP PAIN IN CHILDREN (TABLE 3–9)

TABLE 3–9 Hip Pain in Children

	CONGENITAL HIP DISLOCATION	SCFE	LCPD
Definition	• Dislocated hip at birth	• Femoral head may slip, displacing it medially and posteriorly in relation to the shaft of the femur at the level of the proximal femoral epiphysis	• Idiopathic AVN of the femoral head
Present	• Birth	• Males:females 2:1 • 13- to 16-year-old males • 11- to 13-year-old females • Bilateral involvement: 30%–40%	• Onset: 2–12 years old; if onset is >12 years old, it is considered AVN not LCPD • Boys >> girls • Majority—unilateral
Etiologic Factors	• First born—tight uterine and abdominal musculature of mother • Inhibits fetal movement • Breech position • Left hip > right • Hormonal factors • More common in Whites	• Strain on the growth plate • During its growth spurt secondary to increased weight • 🕮 Endocrinopathies linked with SCFE • Hypothyroidism—most common • Growth hormone abnormalities • Down syndrome	• Bone age low for age results in short stature • Etiology unknown • Linked with hypothyroid abnormality
Signs and Symptoms	• Barlow's test—dislocation; start with hip in flexion and abduction, then the femoral head is dislocated on hip flexion and adduction	• Pain and altered gait • Pain in the groin, medial thigh, and knee • Chronic slip—most common • Loss of internal rotation—when the hip is flexed, it rolls into external rotation	• Mild to intermittent or no pain • Stiffness • Painless limp → antalgic gait • Hip flexion contracture—use Thomas's test

(*continued*)

TABLE 3–9 Hip Pain in Children (*continued*)

	CONGENITAL HIP DISLOCATION	SCFE	LCPD
Signs and Symptoms (*cont.*)	• Ortolani's test—relocation; hip is relocated on hip flexion and abduction	• Acute slip • Trauma, sudden onset of pain on weight-bearing • Acute or chronic chondrolysis • Erosion and degeneration of the cartilage inflaming the synovial membrane on activity	• Limitations in abduction and internal rotation • Disuse atrophy • Short stature
Radiologic Findings	• Not useful until 6 weeks • Negative finding on x-ray does not rule out a dislocation • Proximal and lateral migration of the femoral head from the acetabulum • Acetabular dysplasia • Delayed ossification	• Must obtain AP and frog-leg views of the hip/pelvis • Grading based on degree of displacement of the epiphysis – Grade I: <33% – Grade II: 33%–50% – Grade III: >50%	• Plain films and frog-leg views of the hip/pelvis • Chronological sequence: (1) Growth arrest—avascular stage (2) Subchondral fracture—"crescent sign" (3) Resorption (4) Reossification (5) Healed
Treatment	• Goal—return the hip to its normal position until there is resolution of the pathologic changes. • Closed reduction—<6 months • Position devices: Triple diapers, Frejka pillows, splints: Craig, Von • Rosen-Pavlik harness: Allows hip motion within the safe zone while maintaining abduction • Traction, casting, surgical	• Bedrest-weight relief • Prevention of further displacement • Surgery is the preferred method of treatment—Knowles pinning • Nonsurgical—traction, body casts, hormonal therapy	1. Eliminate hip pain 2. Restoration of motion 3. Prevent femoral head collapse Containment techniques: • Permit weight-bearing of the femoral head to assist healing and remodeling • >6 years old • Abduction braces, Petrie casts, Toronto brace, Salter stirrup • Surgical—epiphysiodesis, valgus osteotomy
Complications	• AVN	• Chondrolysis • AVN • OA	• AVN

AVN, avascular necrosis; AP, anterior-posterior; LCPD, Legg–Calvé–Perthes disease; OA, osteoarthritis; SCFE, slipped capital femoral epiphysis.

Sources: From Jensen HP, Steinke MS, Mikkelsen SS, Thomsen PB. Hip physiolysis: bilaterally in 62 cases followed for 20 years. *Acta Orthop Scand*. 1990;61(5):419–420. doi:10.3109/17453679008993553; Koop S, Quanbeck D. Three common causes of childhood hip pain. *Pediatr Clin North Am*. 1996;43(5):1053–1066. doi:10.1016/S0031-3955(05)70450-8; Skaggs DL, Tolo VT. Legg-Calvé-Perthes disease. *J Am Acad Orthop Surg*. 1996;4(1):9–16. doi:10.5435/00124635-199601000-00002; Wilder RT. Reflex sympathetic dystrophy in children and adolescents: differences from adults. In: Janig W, Stanton-Hicks M, eds. *Reflex Sympathetic Dystrophy: A Reappraisal. Progress in Pain Research and Management Series*, vol 6. IASP Press; 1996.

Thomas's Test
Detects flexion contractures of the hip and evaluates range of hip flexion
1. Patient supine, hand under lumbar region to stabilize, and flex hip.
2. Notice where patient's back touches hand.
3. Flex one hip, then the other as far as possible.
4. Have patient hold one leg on chest, lower the other leg flat.

Indicators of fixed flexion contracture/deformity:
1. Hip does not extend fully.
2. Patient rocks forward, lifts thoracic region, or arches back to reform lordosis.

Acute Transient Synovitis
- Most common cause of hip pain in kids (preadolescents)
- Self-limiting with good outcome
- "Good outcome"

Causes of Avascular Necrosis
"PLASTIC RAGS"

P—pancreatitis	C—caisson disease, collagen vascular disease
L—lupus	R—radiation
A—alcohol	A—amyloid
S—steroids	G—Gaucher's disease
T—trauma	S—sickle cell
I—idiopathic, infection	

FIBROMYALGIA SYNDROME

CLINICAL FEATURES

- Diffuse aching stiffness and fatigue with multiple tender points in specific areas (Figure 3–5)
  - Headaches
  - Neck and upper trapezius discomfort
  - UE paresthesias
  - Fatigue—lack of sleep
  - Brain fog
  - Hyperesthesia, allodynia
- Females > males
- Age—20 to 60 years old
- May experience morning stiffness but it varies throughout the day
- Triggers may exacerbate symptoms:
  - Physical activity
  - Inactivity
  - Sleep disturbance
  - Emotional stress
- May be associated with irritable bowel syndrome, RA, Lyme disease, or hyperthyroidism

1990 AMERICAN COLLEGE OF RHEUMATOLOGY CRITERIA FOR FIBROMYALGIA SYNDROME

- Widespread pain: Pain found in all four quadrants of the body, the left and right sides of the body, as well as above and below the waist
  - Axial involvement—cervical, anterior chest, thoracic, and low back

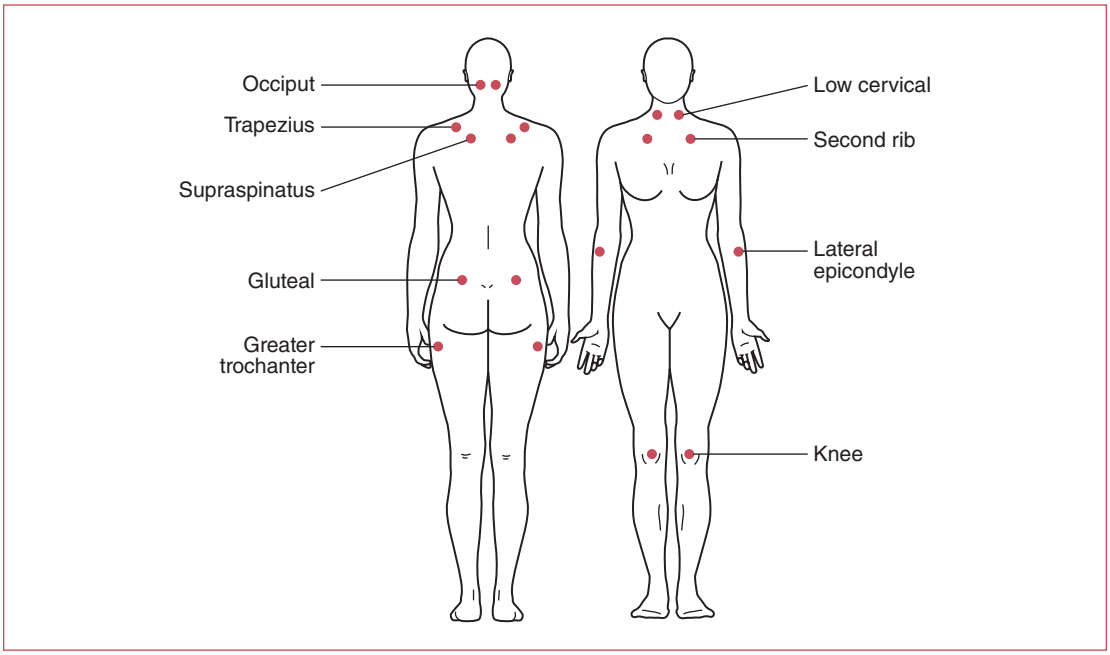

FIGURE 3–5 Fibromyalgia: Location of specific tender points.

- Pain in 11 to 18 tender points (Figure 3–5)
  - Bilateral involvement
  - Occipital, lower cervical, trapezius, supraspinatus, second rib, lateral epicondyle, gluteal, greater trochanter, and knee

2010/2011 American College of Rheumatology Diagnostic Criteria for Fibromyalgia

- In 2010, the ACR published *nontender point diagnostic criteria* for fibromyalgia as an alternative diagnostic method to the 1990 criteria (Wolfe et al., 2010). The 2010 criteria was based on:
  - A widespread pain index score (WPI)
  - A symptom severity score (SSS) that includes fatigue, waking unrefreshed, cognitive symptoms, and somatic symptoms—headaches, abdominal pain/cramps, and depression
  - Symptoms present consistently for ≥3 months
  - Rule out other disorders that could cause the pain symptoms

2016 Revisions to the 2010/2011 American College of Rheumatology Fibromyalgia Diagnostic Criteria

- 🕮 Fibromyalgia diagnostic criteria was subsequently updated in 2016 to include the use of a generalized pain criterion to ensure that regional pain syndromes were not mistakenly included as a fibromyalgia diagnosis. Additionally, the 2016 criteria firmly established fibromyalgia as a separate diagnosis irrespective of other diagnoses (Wolfe et al., 2016).
- WPI ≥7 and SSS score ≥5 OR WPI 4 to 6 and SSS score ≥9
- Generalized pain—defined as pain in at least four of five regions (left upper region, right

2016 ACR Fibromyalgia Diagnosis Criteria

- Widespread pain index (WPI) ≥7 and symptom severity scale (SSS) score ≥5 OR WPI 4–6 and SSS score ≥9
- Generalized pain—defined as pain in at least four of five regions
- Symptoms present for at least 3 months
- A diagnosis of fibromyalgia does not require exclusion of other conditions if other criteria are met

(Wolfe et al., 2016)
ACR, American College of Rheumatology.

upper region, axial region, left lower region, and right lower region). In this definition, jaw, chest, and abdominal pain is not evaluated as part of the generalized pain definition.
- Symptoms have been present at a similar level for at least 3 months.
- A diagnosis of fibromyalgia is valid irrespective of other diagnoses. A diagnosis of fibromyalgia does not exclude the presence of other clinically important illnesses.

TREATMENT OF FIBROMYALGIA SYNDROME

- Patient education and reassurance
- Combination therapy often more effective
- Medications:
  - Pregabalin (Lyrica), duloxetine (Cymbalta), and milnacipran (Savella) are the only Food and Drug Administration-approved medications to treat fibromyalgia.
  - Tricyclic antidepressants (amitriptyline, nortriptyline)
  - Muscle relaxants (cyclobenzaprine, tizanidine)
  - Tramadol
- Biofeedback, tender point injections
- Acupuncture
- Low-impact, graded aerobic exercise
- Tai Chi
- Cognitive behavioral therapy
- Aquatic based exercises

FIBROMYALGIA VERSUS MYOFASCIAL PAIN SYNDROME AND CHRONIC FATIGUE SYNDROME

- Fibromyalgia should be differentiated from myofascial pain syndrome and chronic fatigue syndrome.

Myofascial Pain Syndrome

- Characterized by local pain and tender points in the muscles with taut bands and trigger points. Resolve with PT and local treatment but may recur.
- Fatigue, morning stiffness uncommon

Chronic Fatigue Syndrome

- Disabling fatigue for at least 6 months
- Often preceded by a viral syndrome

COMPLEX REGIONAL PAIN SYNDROME

- Also see the Complex Regional Pain Syndrome Type I section in Chapter 1, Stroke, and Chapter 11, Pain Medicine, for a more detailed discussion.
- Complex regional pain syndrome (CRPS) type I:
  - Formerly known as:
    - Reflex sympathetic dystrophy
    - Sudeck's atrophy
    - Algodystrophy
    - Shoulder hand syndrome
  - Occurs after a traumatic injury and without a specific nerve injury
- CRPS type II is also known as causalgia and is seen after a specific nerve injury.

CHARACTERISTICS

- Limb pain, swelling, and autonomic dysfunction
- Most commonly caused by minor or major trauma

CLINICAL FEATURES

- Pain, deep burning sensations exacerbated by movement:
  - Allodynia—pain induced by a nonnoxious stimulus
  - Hyperalgesia—lower pain threshold and enhanced pain perception
  - Hyperpathia
- Local edema and vasomotor changes:
  - Extremity warm, red, and dry initially
  - Later becomes cool, mottled, and cyanotic
- Muscle weakness
- Dystrophic changes:
  - Thin, shiny skin, and brittle nails

🕮 CLINICAL STAGES

1. **Acute:** Few weeks to 6 months:
   - Allodynia, hyperpathia, hypersensitivity, swelling, and vasomotor changes
   - Increased blood flow creating temperature and skin-color changes
   - Hyperhidrosis
2. **Dystrophic:** 3 to 6 months:
   - Persistent pain, disability, and atrophic skin changes
   - Decreased blood flow, decreased temperature
   - Hyperhidrosis
3. **Atrophic:**
   - Atrophy and contractures
   - Skin glossy, cool, and dry

🕮 RADIOGRAPHIC FINDINGS

1. Plain radiographs:
   - Sudeck's atrophy—patchy osteopenia, ground-glass appearance
2. Triple-phase bone scan:
   - First two phases nonspecific
   - Third-phase bone scan—abnormal, with enhanced uptake in the periarticular structures

TREATMENT

1. Immediate mobilization—passive and active ROM, massage, contrast baths, transcutaneous electrical stimulation
2. 🕮 Advise patients to continue activities as tolerated to avoid disuse atrophy.
3. Pain control
   - See Chapter 11, Pain Medicine
   - Few clinical trials have been completed specifically in regard to CRPS; the focus is on treating the barriers to a patient's participation in mobilization activities.
4. Inflammation—corticosteroids, initial dose 60 to 80 mg four times a day dosing for 2 weeks, then gradual tapering the next 2 weeks
5. Cervical sympathetic ganglia block for the upper extremities, lumbar ganglion block for the lower extremities
6. Surgical sympathectomy—if block is beneficial but transient

TABLE 3–10 Complex Regional Pain Syndrome in Children and Adolescents Versus Adults

	CHILDREN/ADOLESCENTS	ADULTS
Site	Lower extremity	Upper extremity
Spontaneous pain	Common	Common
Allodynia	Most patients	Most patients
Sex ratio	4:1 female > male	Mixed
Three-phase bone scan	• Mixed results: Used to rule out other pathology • See decreased uptake of the extremity—decreased atrophic changes • Occasionally normal • Will have increased uptake normally secondary to bone growth	Increased uptake in the third-phase bone scan of the affected extremity
Treatment	• Physical therapy alone • Noninvasive—TENS, biofeedback • Meds—tricyclic antidepressant • Blocks more common in the upper extremity	Sympathetic blocks
Prognosis	Good	Poor

TENS, transcutaneous electrical stimulation.
Source: From Wilder RT. Reflex sympathetic dystrophy in children and adolescents: differences from adults. In: Janig W, Stanton-Hicks M, eds. *Reflex Sympathetic Dystrophy: A Reappraisal. Progress in Pain Research and Management Series, vol 6*. IASP Press; 1996, with permission.

🕮 SYMPATHETICALLY MEDIATED COMPLEX REGIONAL PAIN SYNDROME

Four tests are used to determine if the pain is sympathetically mediated. The first two are used more commonly.

1. Sympathetic block with local anesthetic:
   - Local anesthetic is injected at the stellate ganglion (UE) or the lumbar paravertebral ganglion (LE). If relief, then suspect sympathetic etiology.
   - 🕮 A positive response to a stellate ganglion block includes ipsilateral Horner's syndrome, anhidrosis, conjunctival injection, nasal congestion, vasodilation, and an increased skin temperature. A positive Horner's syndrome by itself after the block does not equate to a successful sympathetic block (Elias, 2000). See Chapter 11, Pain Medicine, for a more detailed description.
2. Guanethidine test:
   - Injection of guanethidine into the extremity distal to a suprasystolic cuff. The test is positive if the pain is reproduced after injection and is immediately relieved after the cuff is released.
3. Phentolamine test:
   - IV phentolamine will reproduce the pain.
4. Ischemia test:
   - Inflation of the suprasystolic cuff decreases the pain.

TENDON DISORDERS

🕮 DUPUYTREN'S CONTRACTURE (FIGURE 3–6)

- Abnormal fibrous hyperplasia and contracture of the palmar fascia, causing a flexion contracture at the MCP and PIP joints
- More common in White men approximately 50 to 70 years of age

- Associated with epilepsy, pulmonary TB, alcoholism, and diabetes mellitus (Snider, 1997)

Mechanism

- The palmar fascia is a continuation of the palmaris longus tendon attaching to the sides of the PIP and middle phalanges as well as to the skin.
- Fibromatosis of the palmar fascia and contracture of the fibrous bands that develop into nodules can lead to the development of a finger flexion contracture and skin dimpling.

Clinical Features

- Painless thickening of the palmar surface and underlying fascia
- Most commonly at the fourth and fifth digits

Treatment

- Nonoperative—PT, injections (steroid, anti-TNF agents, collagenase), percutaneous needle fasciotomy (Denkler et al., 2022; Smeraglia et al., 2016)
- Modalities—heating, stretching, ultrasound
- Surgical—fasciotomy, amputation

TRIGGER FINGER (STENOSING FLEXOR TENOSYNOVITIS; FIGURE 3–7)

- Thickening of the flexor tendon sheath causes increased friction through normal movement.
- A nodule in the tendon sheath may develop, causing the tendon to "catch" at the A1 pulley system and not glide through, limiting finger movement.
- A locking ("catching") or clicking sensation is felt when the nodule passes though the tendon sheath of the pulley system.
- When the finger is flexed, the nodule moves proximally and reextension is prevented.

🕮 MALLET FINGER (FIGURE 3–8)

- Most common extensor tendon injury (Snider, 1997)
- Rupture of the extensor tendon into the distal phalanx secondary to forceful flexion
- The DIP drops remain in a flexed position and cannot be actively extended.
- Treatment: DIP splint immobilizes the distal phalanx in hyperextension:
  - Acute—6 weeks
  - Chronic—12 weeks
- Surgical indications: Poor healing, volar subluxation, avulsion of greater than one third of the bone

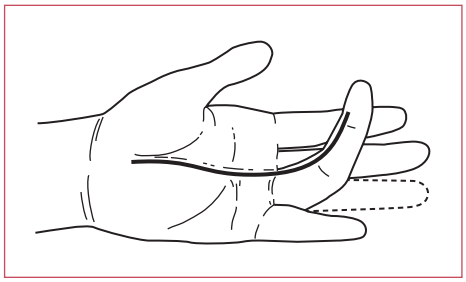

FIGURE 3–6 Dupuytren's contracture.
Source: From Snider RK, ed. *Essentials of Musculoskeletal Care*. American Academy of Orthopaedic Surgeons; 1997, with permission.

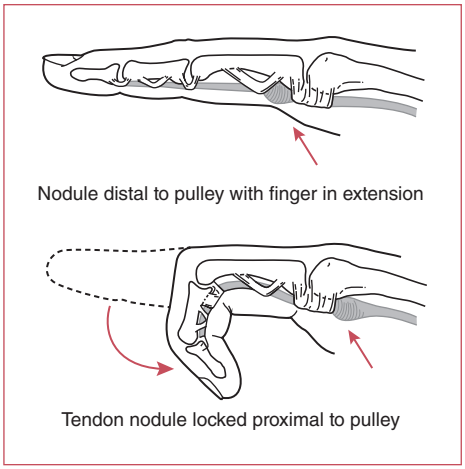

FIGURE 3–7 Trigger finger: With the finger in extension, the nodule is distal to the pulley. When the finger is flexed, the tendon locks proximal to the A1 pulley.
Source: From Snider RK, ed. *Essentials of Musculoskeletal Care*. American Academy of Orthopaedic Surgeons; 1997, with permission.

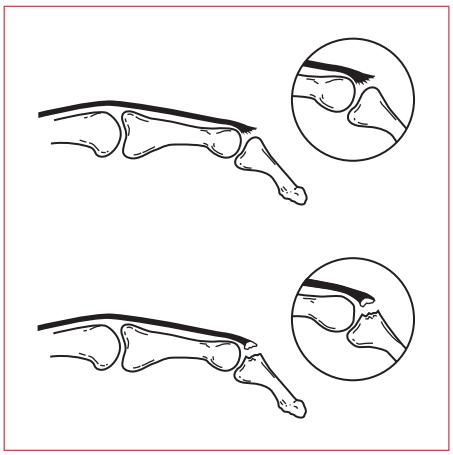

FIGURE 3–8 Mallet finger caused by:
Top: Rupture of the extensor tendon at its insertion. *Bottom*: Avulsion of a portion of the distal phalanx.

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MUSCULOSKELETAL MEDICINE

4

UPPER EXTREMITIES—David P. Brown, DO • Eric D. Freeman, DO • Sara J. Cuccurullo, MD • Sagar Parikh, MD • Laurent Delavaux, MD • Ian B. Maitin, MD, MBA • Alexander Shustorovich, DO • Michael Chung, MD

LOWER EXTREMITIES—David P. Brown, DO • Eric D. Freeman, DO • Sara J. Cuccurullo, MD • Sagar Parikh, MD • Laurent Delavaux, MD • Ian B. Maitin, MD, MBA • Craig Van Dien, MD • Michael Galibov, DO

SPINE—Ted L. Freeman, DO • Eric D. Freeman, DO

UPPER EXTREMITIES: THE SHOULDER REGION

FUNCTIONAL ANATOMY

Ranges of Motion of the Shoulder (Figure 4–1)

- Shoulder flexion: 180 degrees
- Shoulder extension: 60 degrees
- Shoulder abduction: 180 degrees:
  - Shoulder abduction of 120 degrees is seen with the thumb pointed down.

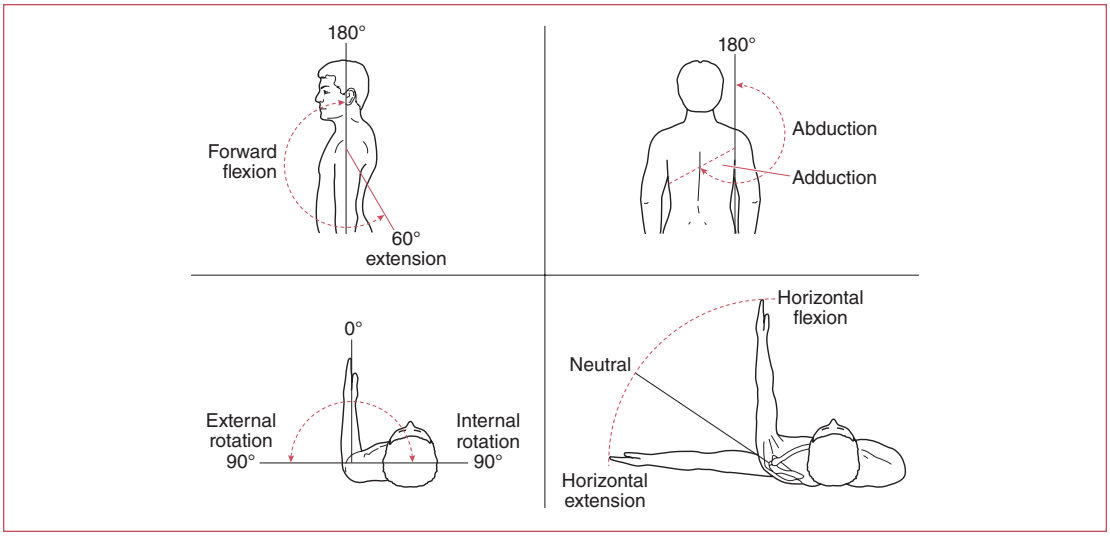

FIGURE 4–1 Shoulder range of motion.

- Shoulder adduction: 60 degrees
- Shoulder internal rotation: 90 degrees (with arm abducted)
- Shoulder external rotation: 90 degrees (with arm abducted)

Shoulder Motions

SHOULDER FLEXION (FIGURE 4–2)

SHOULDER FLEXORS	SPINAL INNERVATION	PERIPHERAL INNERVATION
Anterior deltoid	C5, C6	Axillary nerve from posterior cord
Pectoralis major, clavicular portion	C5, C6, C7, C8, T1	Medial and lateral pectoral nerves
Biceps brachii	C5, C6	Musculocutaneous nerve from lateral cord
Coracobrachialis	C5, C6	Musculocutaneous nerve from lateral cord

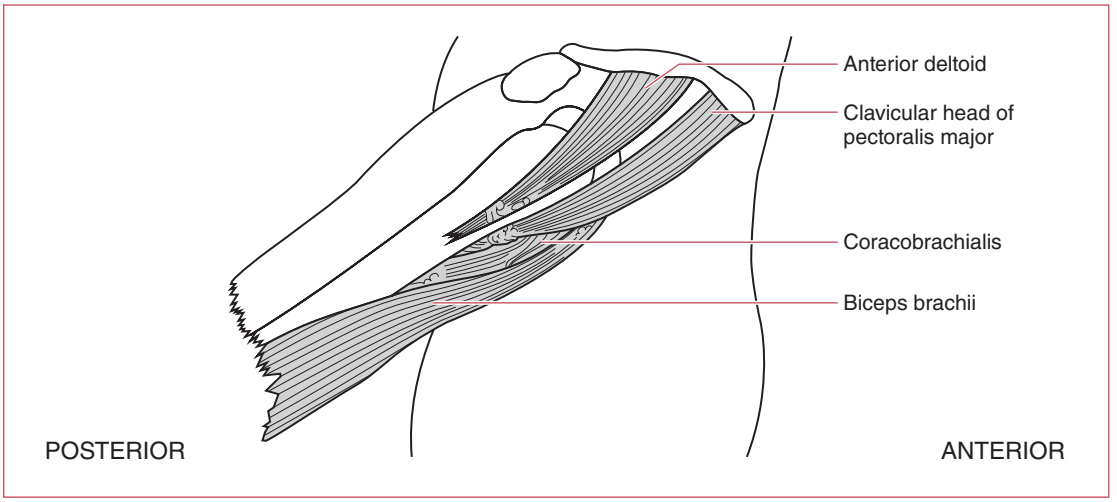

FIGURE 4–2 Shoulder flexors (lateral view). Please note arm is shown in extension to better appreciate flexor muscle attachments.

SHOULDER EXTENSION (FIGURE 4–3)

SHOULDER EXTENSORS	SPINE INNERVATION	PERIPHERAL INNERVATION
Posterior deltoid	C5, C6	Axillary nerve from posterior cord
Latissimus dorsi	C6, C7, C8	Thoracodorsal nerve from posterior cord
Teres major	C5, C6	Lower subscapular nerve from posterior cord
Triceps, long head	C6, C7, C8	Radial nerve from posterior cord
Pectoralis major, sternocostal portion	C5, C6, C7, C8, T1	Medial and lateral pectoral nerves

SHOULDER ABDUCTION (FIGURE 4–4)

SHOULDER ABDUCTORS	SPINE INNERVATION	PERIPHERAL INNERVATION
Middle deltoid	C5, C6	Axillary nerve from posterior cord
Supraspinatus	C5, C6	Suprascapular nerve from upper trunk

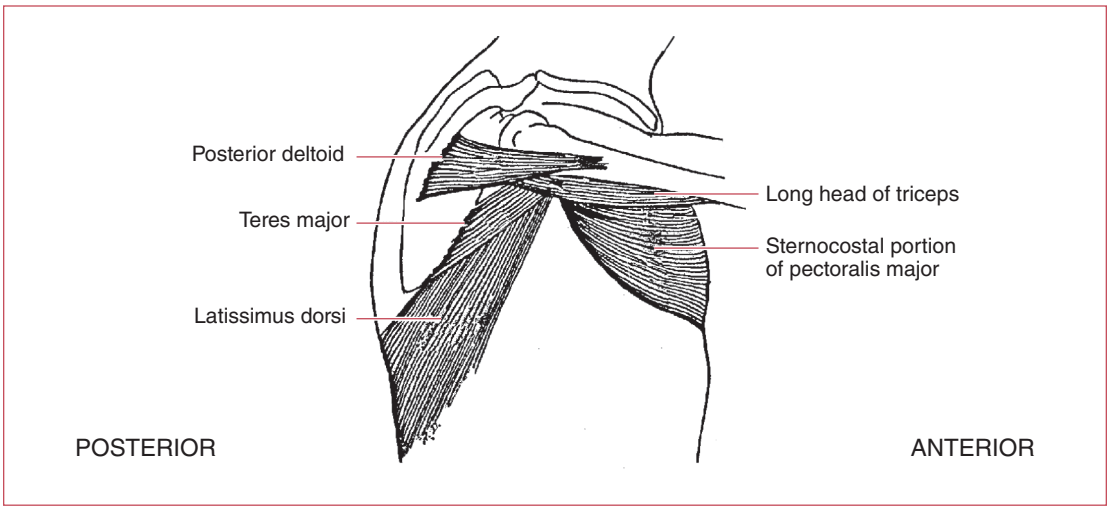

FIGURE 4–3 Shoulder extensors (lateral view). Please note arm is shown flexed at the shoulder to better appreciate extensor muscle attachments.

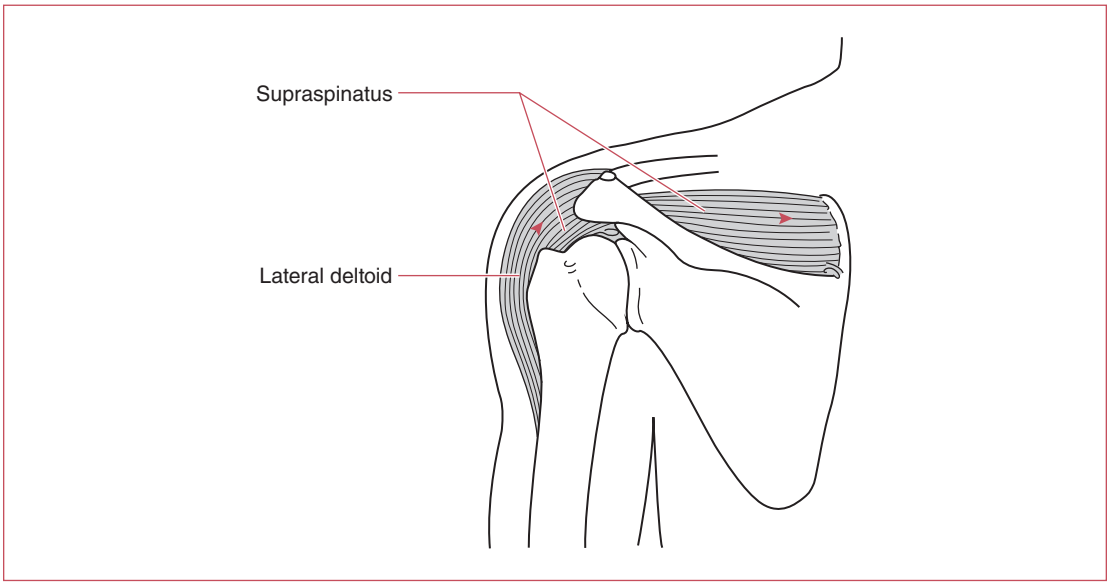

FIGURE 4–4 Shoulder abductors (posterior view).

SHOULDER ADDUCTION (FIGURE 4–5)

SHOULDER ADDUCTORS	SPINE INNERVATION	PERIPHERAL INNERVATION
Pectoralis major	C5, C6, C7, C8, T1	Medial and lateral pectoral nerves
Latissimus dorsi	C6, C7, C8	Thoracodorsal nerve from posterior cord
Teres major	C5, C6	Lower subscapular nerve from posterior cord
Coracobrachialis	C5, C6, C7	Musculocutaneous nerve from lateral cord
Infraspinatus	C5, C6	Suprascapular nerve from upper trunk
Long head of triceps	C6, C7, C8	Radial nerve from posterior cord
Anterior and posterior deltoid	C5, C6	Axillary nerve from posterior cord

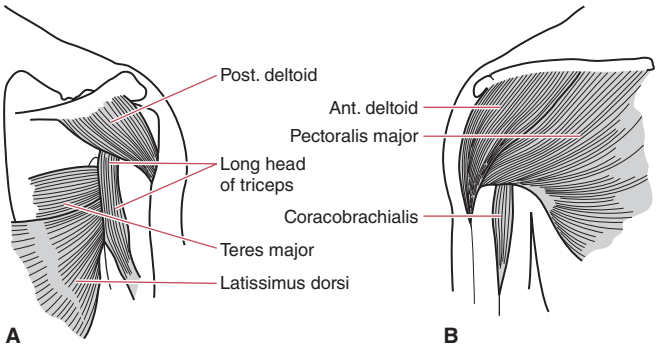

FIGURE 4–5 Shoulder adductors. (A) Posterior view. (B) Anterior view.

SHOULDER INTERNAL ROTATION (FIGURE 4–6)

SHOULDER INTERNAL ROTATORS	SPINE INNERVATION	PERIPHERAL INNERVATION
Subscapularis	C5, C6	Upper and lower subscapular nerves from posterior cord
Pectoralis major	C5, C6, C7, C8, T1	Medial and lateral pectoral nerves
Latissimus dorsi	C5, C6	Thoracodorsal nerve from posterior cord
Anterior deltoid	C5, C6	Axillary nerve from posterior cord
Teres major	C5, C6	Lower subscapular nerve from posterior cord

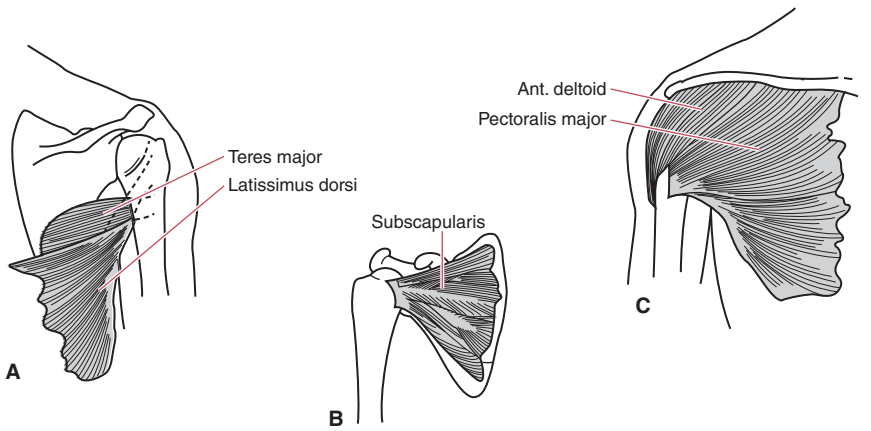

FIGURE 4–6 Shoulder internal rotators. (A) Posterior view. (B and C) Anterior views.

SHOULDER EXTERNAL ROTATION (FIGURE 4–7)

SHOULDER EXTERNAL ROTATORS	SPINE INNERVATION	PERIPHERAL INNERVATION
Infraspinatus	C5, C6	Suprascapular nerve from upper trunk
Teres minor	C5, C6	Axillary nerve from posterior cord
Deltoid, posterior portion	C5, C6	Axillary nerve from posterior cord
Supraspinatus	C5, C6	Suprascapular nerve from upper trunk

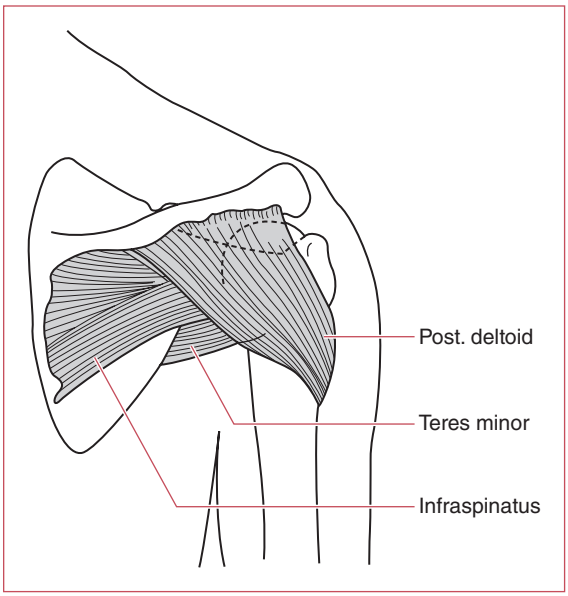

FIGURE 4–7 Shoulder external rotators (posterior view).

The Shoulder–Girdle Complex

THE GLENOHUMERAL JOINT

- The glenohumeral (GH) joint consists of a ball-and-socket type synovial joint.
- Main components of the GH joint:
  - Glenoid fossa and humerus
  - Labrum
  - Glenohumeral capsule
  - Glenohumeral ligaments
  - Dynamic shoulder stabilizers
  - Static shoulder stabilizers
- 🕮 Arm abduction is achieved through GH and scapulothoracic joint motion.
- 🕮 Balance exists between the GH and scapulothoracic joint during arm abduction.
  - There are two degrees of GH motion for every one degree of scapulothoracic motion during arm abduction (120 degrees of GH motion to 60 degrees of scapulothoracic motion).
  - The scapulothoracic motion allows the glenoid to rotate and permits GH abduction without acromial impingement.

GLENOID FOSSA (FIGURE 4–8)

- Lateral aspect of the scapula that articulates with the humerus
- Approximately 30% of the humeral head articulates with the glenoid fossa.

LABRUM (FIGURE 4–8)

- Fibrocartilaginous tissue surrounding the glenoid fossa
- Serves as an attachment site for the GH ligaments and tendons as well as the shoulder capsule
- Prevents anterior and posterior humeral head dislocation
- Deepens the glenoid fossa and increases overall contact of the humeral head with the glenoid by 70%

GLENOHUMERAL CAPSULE

- The capsule arises from the labrum, covers the entire head of the humerus, and attaches to the neck of the humerus.
- The capsule thickens anteriorly to form the GH ligaments.

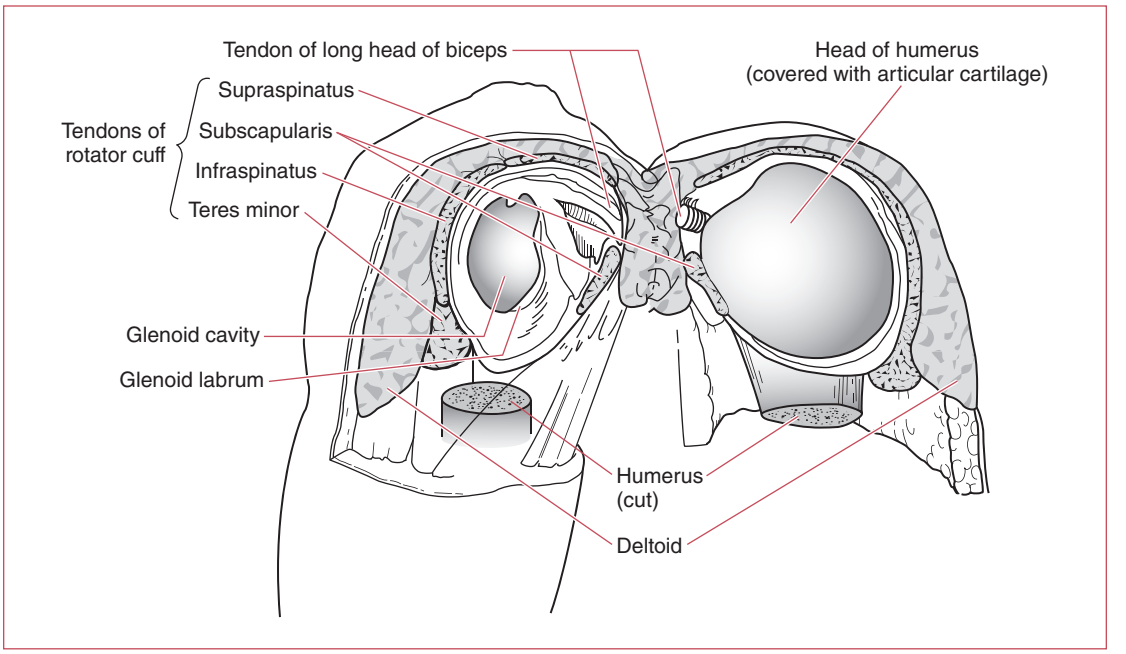

FIGURE 4–8 The glenoid labrum and glenoid fossa (lateral view).

GLENOHUMERAL LIGAMENTS (FIGURE 4–9)

- These ligaments arise from folds of the anterior portion of the GH capsule and attach to the glenoid to reinforce the shoulder capsule and joint.
- They provide stability and prevent translation of the head of the humerus from the glenoid fossa.

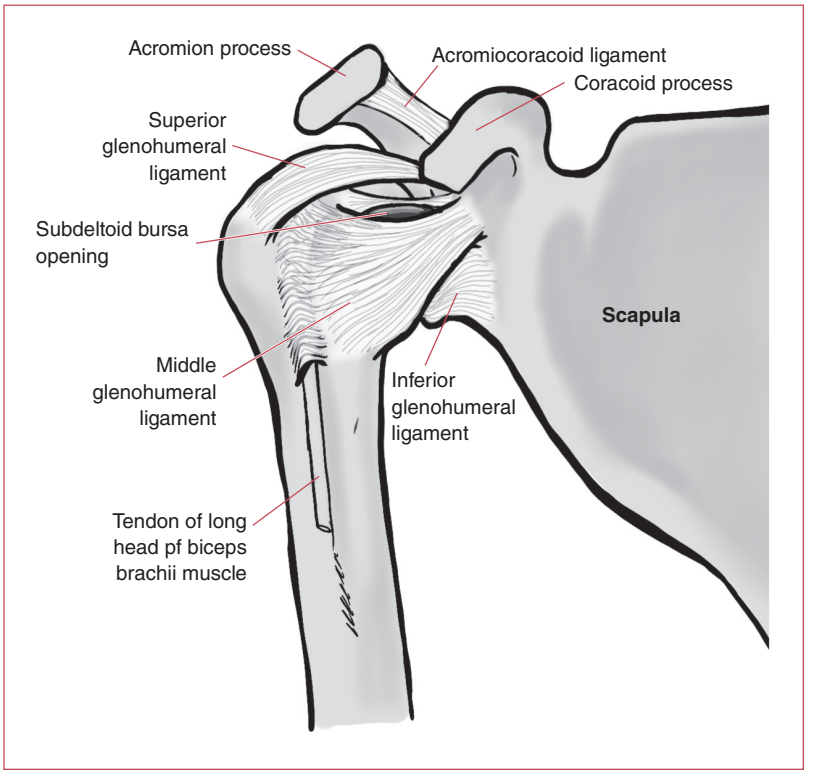

FIGURE 4–9 The GH ligaments (anterior view) depict a distinct Z-pattern formed by the superior GH ligament, the middle GH ligament, and the inferior GH ligament. *Note:* The opening for the subdeltoid bursa is variable. GH, glenohumeral. *Source:* Illustration by Sagar Parikh, MD.

- They are composed of three segments, all of which are located on the anterior aspect of the humeral head
  1. **Superior** GH ligament
     - Prevents translation in the inferior direction
     - This, along with the middle GH ligament, provides stability of the shoulder from 0 to 90 degrees of abduction.
  2. **Middle** GH ligament
     - Prevents anterior shoulder translation
  3. **Inferior** GH ligament
     - The primary anterior ligament stabilizer above 90 degrees

Shoulder Joint Stability

DYNAMIC STABILIZERS

- Surround the humeral head and help to approximate it into the glenoid fossa
- Rotator cuff muscles: "S.I.T.S." (**Figures 4–10, 4–11, and 4–19**):
  - Supraspinatus
  - Infraspinatus
  - Teres minor
  - Subscapularis
- Long head of the biceps tendon, deltoid, and teres major, latissimus dorsi
- Scapular stabilizers (e.g., trapezius, serratus anterior) play a supporting role in stabilizing the GHJ during shoulder range of motion (ROM)

STATIC STABILIZERS

- These include the glenoid, the labrum, the shoulder capsule, and the GH ligament.

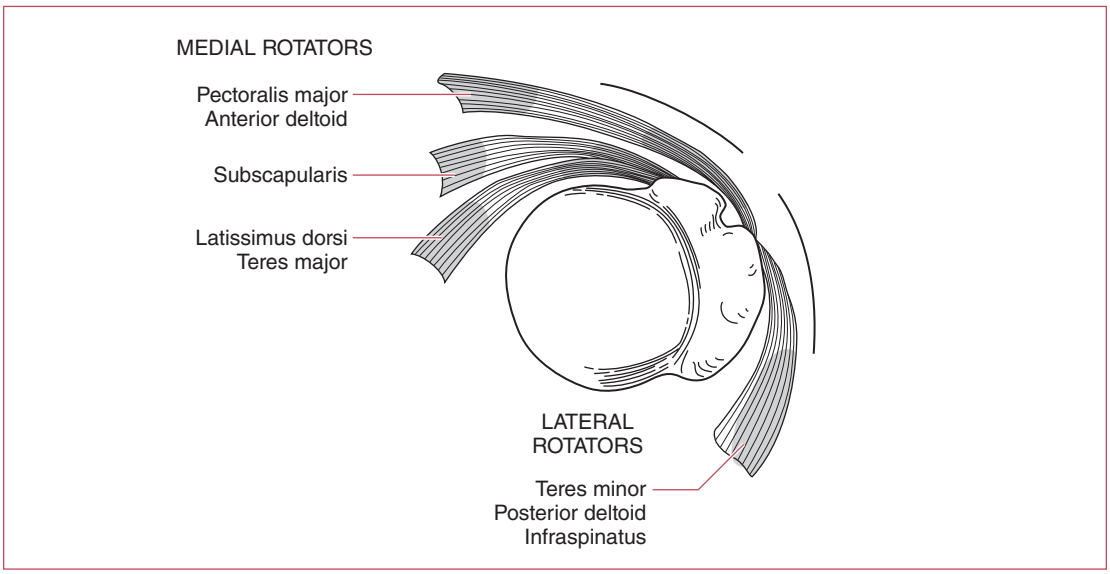

FIGURE 4–10 Right arm superior view: Medial rotator; lateral rotators. This diagram depicts the relation of the rotators to the upper end of the humerus.

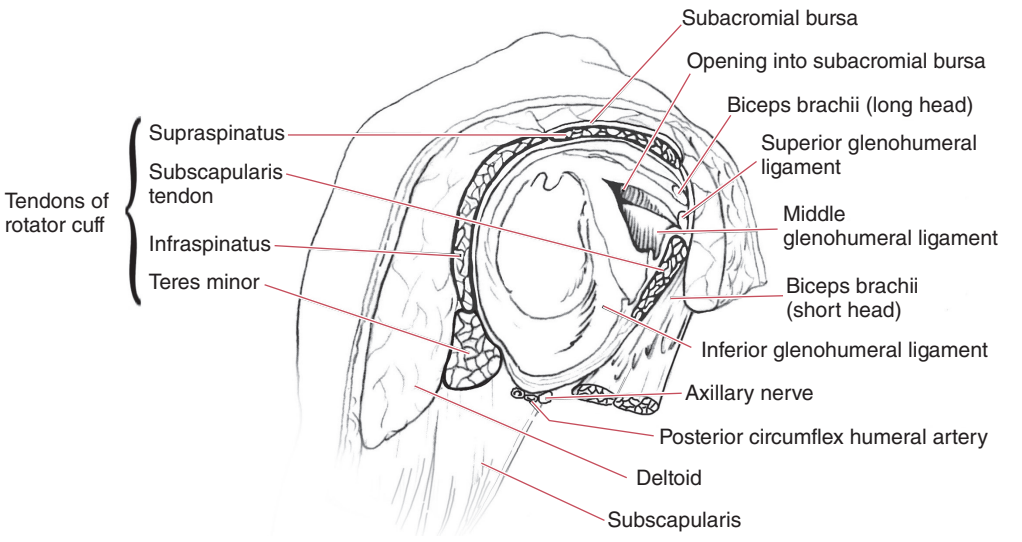

FIGURE 4–11 Right glenoid cavity of the scapula as viewed from the anterolateral aspect. Note the four short rotator cuff muscles (teres minor, infraspinatus, supraspinatus, and subscapularis).

SHOULDER DISORDERS

ACROMIOCLAVICULAR JOINT INJURIES

General

ACROMIOCLAVICULAR JOINT (FIGURE 4–12)

- Gliding joint that anchors the clavicle to the scapula
- Articular disc between the two joint surfaces

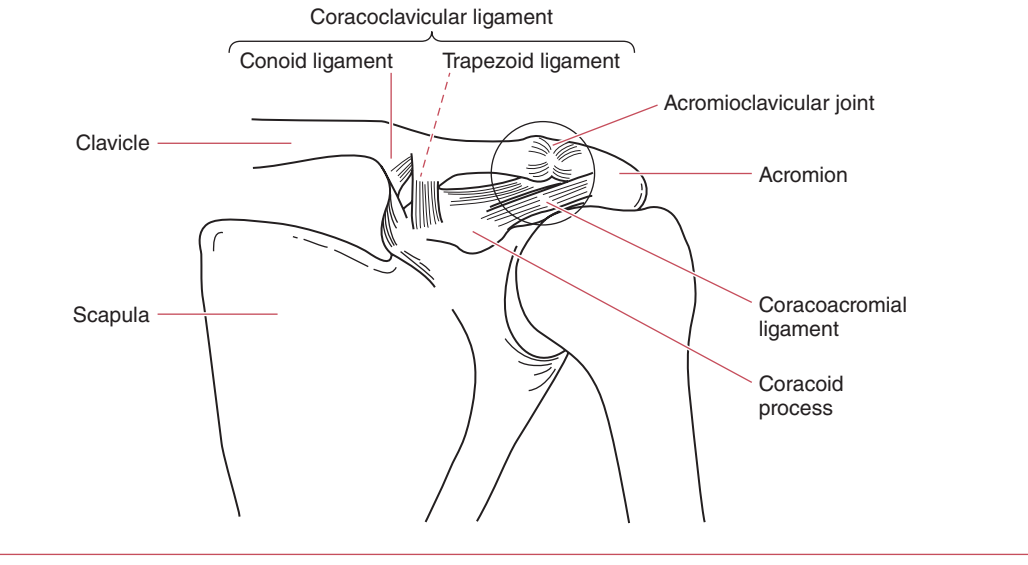

FIGURE 4–12 Anterior view of the acromioclavicular joint. Note the contribution of the coracoacromial ligaments to the inferior acromioclavicular joint capsule.

ACROMIOCLAVICULAR LIGAMENTS

- The **acromioclavicular (AC) ligament** connects the distal end of the clavicle to the acromion, providing horizontal stability.
- The **coracoclavicular (CC) ligament** connects the coracoid process to the clavicle and anchors the clavicle to the coracoid process, preventing vertical translation of the clavicle. It is made up of the conoid and trapezoid ligaments.
- The **coracoacromial ligament** connects the coracoid process to the acromion.

MECHANISM OF INJURY

- A direct impact to the shoulder
- Falling on an outstretched arm

Classification of Acromioclavicular Joint Separations (Table 4–1 and Figure 4–13)

Clinical Features

- Patients generally complain of tenderness over the AC joint with palpation and ROM.
- AC joint displacement with gross deformity occurs in the later stages and is usually seen in a type III or greater.
- The Rockwood classification of AC joint separations categorizes AC joint injuries:
  - Types I to III are based on the degree of AC and CC ligament injury and sequential displacement of the AC joint.
  - Types IV to VI are expanded classifications that include the direction of the displaced clavicle.

TABLE 4–1 Rockwood Classification of AC Joint (Shoulder) Separations

GRADING	AC LIGAMENT	CC LIGAMENT	AC JOINT/CLAVICLE DISPLACEMENT	EXAM
Type I	Sprained	Intact	AC joint intact No clavicular displacement	AC joint tenderness without instability, soft tissue swelling
Type II	Complete tear	Sprained	AC joint disrupted, slight widening of interval Mildly elevated but not above the superior border of the acromion	AC joint tenderness with horizontal instability
Type III	Complete tear	Ruptured	Dislocated, widened interval up to 100% Clavicle elevated above the superior border of the acromion but CC distance 25%–100% of contralateral side	
Type IV	Complete rupture	Complete rupture	Clavicle displaced posteriorly through trapezius	Skin tenting
Type V	Complete rupture	Complete rupture	CC distance more than 100% of contralateral side	Severe shoulder droop that does not improve with shrug
Type VI (rare)	Complete rupture	Complete rupture	Distal clavicle inferior to coracoid	

AC, acromioclavicular; CC, coracoclavicular.
Source: Adapted from Gorbaty JD, Hsu JE, Gee AO. Classifications in brief: Rockwood classification of acromioclavicular joint separations. *Clin Orthop Relat Res*. 2017;475(1):283–287. doi:10.1007/s11999-016-5079-6.

PROVOCATIVE TEST FOR ACROMIOCLAVICULAR JOINT IMPINGEMENT

- Cross-chest (horizontal adduction or scarf) test: Passive adduction of the arm across the midline causing joint tenderness

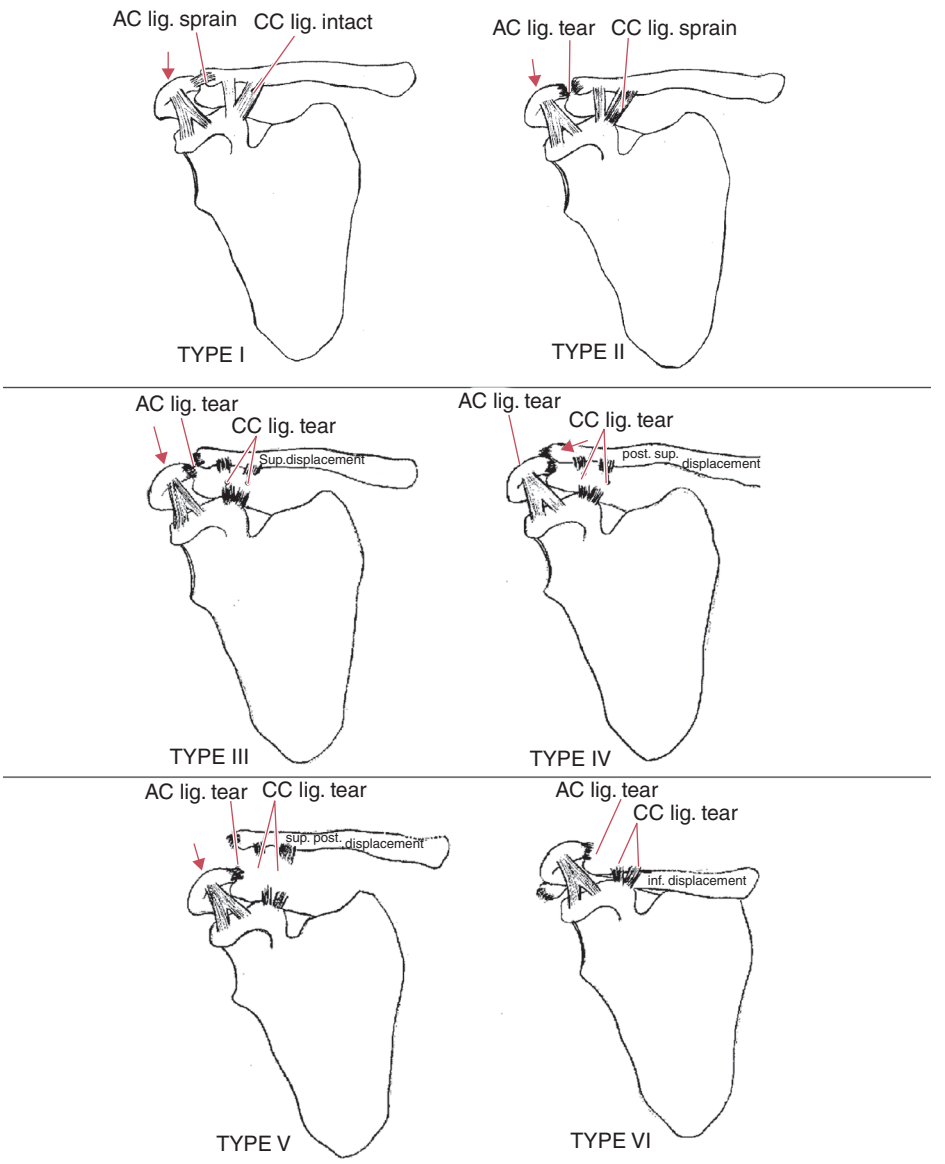

FIGURE 4–13 Classification of AC joint separations (anterior views). (See Table 4–1 for description.) AC, acromioclavicular; CC, coracoclavicular.

Imaging

- Weighted anterior-posterior (AP) radiographs of the shoulders (10 lb.):
  - Type III injuries may show a 25% to 100% widening of the CC space.
  - Type V injuries may show a widening >100%.

Treatment

- Treatment regimens will differ depending on the degree of separation and acuity of injury.

ACUTE ACROMIOCLAVICULAR JOINT INJURIES

- *Types I and II:*
  - Rest, ice, nonsteroidal anti-inflammatory drugs (NSAIDs)
  - Sling for comfort for the first 1 to 2 weeks

  - Avoid heavy lifting and contact sports
  - Shoulder–girdle complex stabilization and strengthening
  - Return to play: When the patient is asymptomatic with full ROM
    - Type I: 2 weeks
    - Type II: 6 weeks
- *Type III:*
  - Treatment is controversial. Conservative or surgical route depends on the patient's need (occupation or sport) for particular shoulder stability.
  - Surgical for those indicated (heavy laborers, athletes)
  - Generally, no functional advantage is seen between the two treatment regimens.
- *Types IV, V, and VI:*
  - Surgery is recommended: Open reduction internal fixation (ORIF) or distal clavicular resection with reconstruction of the CC ligament

CHRONIC ACROMIOCLAVICULAR JOINT INJURIES/PAIN

- Corticosteroid injection
- May require clavicular resection and CC reconstruction

COMPLICATIONS OF ACROMIOCLAVICULAR JOINT INJURIES

- Associated clavicular fractures and dislocations
- Distal clavicle osteolysis: Degeneration of the distal clavicle with associated osteopenia and cystic changes
- AC joint arthritis: May get relief from a corticosteroid injection and conservative rehabilitative care

GLENOHUMERAL JOINT INJURIES

General

GHJ TYPE: BALL AND SOCKET

- *Scapulothoracic motion:*
  - Balance exists between the GH and scapulothoracic joints during arm abduction.
  - The scapulothoracic motion allows the glenoid to rotate and permit GH abduction without acromial impingement.
  - There is a 2:1 GH:scapulothoracic motion accounting for the ability to abduct the arm (60 degrees of scapulothoracic motion to 120 degrees of GH motion).

Classification of GHJ Instability

- **Instability** is a translation of the humeral head with respect to the glenoid fossa. It may result in subluxation or dislocation.
- **Subluxation** is an incomplete separation of the humeral head from the glenoid fossa with immediate reduction.
- **Dislocation** is a complete separation of the humeral head from the glenoid fossa without immediate reduction.

🕮 Direction of Instability

- *Anterior GH instability:*
  - Most common direction of instability is anterior inferior.
  - More common in the younger population and has a high recurrence rate
  - Mechanism: Arm abduction and external rotation
  - Complications may include axillary nerve injury.
- *Posterior GH instability:*
  - Less common than anterior instability
  - May occur as a result of a seizure

  - The patient may present with the arm in the adducted internal rotated position.
  - Mechanism: Landing on a forward flexed adducted arm
- *Multidirectional instability:*
  - Rare with instability in multiple planes
  - The patient may display generalized laxity in other joints.

PATTERNS OF INSTABILITY

- *Traumatic: TUBS*
- *Atraumatic: AMBRI*

TUBS	AMBRI
T—Traumatic shoulder instability	A—Atraumatic shoulder instability
U—Unidirectional	M—Multidirectional instability
B—Bankart lesion	B—Bilateral lesions
S—Surgical management	R—Rehabilitation management
	I—Inferior capsular shift, if surgery

Source: From Rockwood CA, Green DP, Bucholz RW, Heckman JD. *Rockwood and Green's Fractures in Adults*. 4th ed. Lippincott-Raven; 1996.

ASSOCIATED INJURIES WITH SHOULDER DISLOCATIONS

- *Anterior dislocations:*
  - *Bankart lesion* **(Figure 4–14):**
    - Labral tear off the anterior glenoid allows the humeral head to slip anteriorly.
    - Most commonly associated with anterior instability
    - May also be associated with an avulsion fracture off the glenoid rim
  - *Hill–Sachs lesion* **(Figure 4–15):**
    - Compression fracture of the posterolateral humeral head caused by abutment against the anterior rim of the glenoid fossa
    - Associated with anterior dislocations
    - A lesion that accounts for >30% of the articular surface may cause instability.
    - A notch occurs on the posterior lateral aspect of the humeral head.
  - *Posterior dislocations:*
    - Reverse Bankart lesion
    - Reverse Hill–Sachs lesion

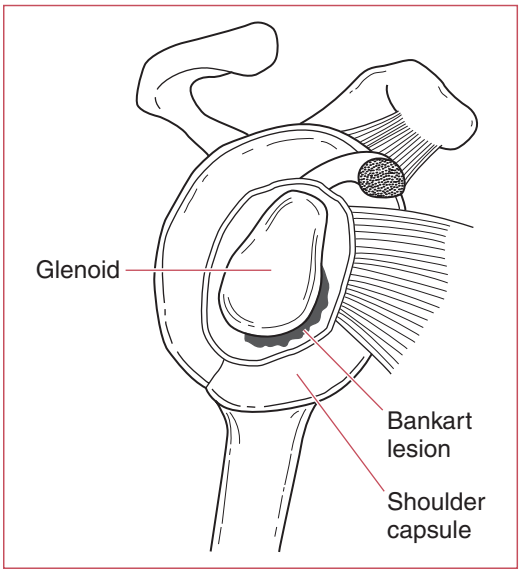

FIGURE 4–14 Bankart lesion.

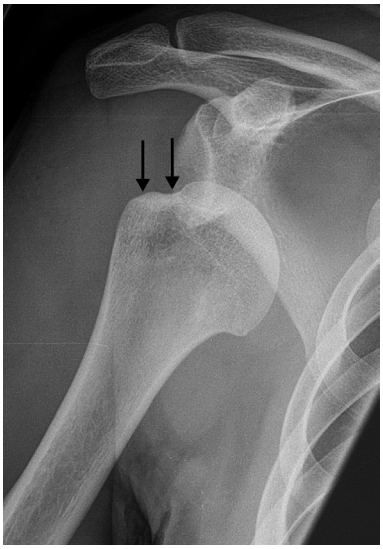

FIGURE 4–15 Hill–Sachs lesion.
Source: Courtesy of Hellerhoff.

Clinical Features

- **Dead arm syndrome:**
  - Symptoms include early shoulder fatigue, pain, numbness, and paresthesias.
  - Shoulder slipping in and out of place most commonly when the arm is placed in the abduction and external rotation ("throwing position")
  - Typically seen in athletes such as pitchers or volleyball players who require repetitive overhead arm motion
- **Laxity exam:** Some patients are double-jointed, which is a lay term for capsular laxity. Ask the patient to touch the thumb against the volar (flexor) surface of the forearm. Patients with lax tissues are more likely than others to be able to voluntarily dislocate the shoulder.

Provocative Tests

FOR ANTERIOR GLENOHUMERAL INSTABILITY:

- **Apprehension test (Figure 4–16):**
  - A feeling of anterior shoulder instability with 90-degree shoulder abduction and external rotation, causing apprehension (fear of dislocation) in the patient
- **Relocation test:**
  - Supine apprehension test with a posterior-directed force applied to the anterior aspect of the shoulder not allowing anterior dislocation. This force relieves the feeling of apprehension.
- **Anterior drawer test:**
  - Passive anterior displacement of the humeral head on the glenoid
- **Anterior load-and-shift test:**
  - Essentially a modified form of the anterior drawer test
  - Humeral head is loaded against the glenoid and then passively displaced anteriorly. Positive if there is reproduction of the patient's symptoms of instability, pain, and crepitation.

FIGURE 4–16 Apprehension test.
Source: Courtesy of JFK Johnson Rehabilitation Institute, 2000.

FOR POSTERIOR GLENOHUMERAL INSTABILITY

- **Jerk test:**
  - Place the arm in 90 degrees of flexion and maximum internal rotation with the elbow flexed 90 degrees. Adduct the arm across the body in the horizontal plane while pushing the humerus in a posterior direction. The patient will jerk away when the arm nears midline to prevent posterior subluxation or dislocation of the humeral head.
- **Posterior drawer test**
- **Posterior load-and-shift test**

FOR MULTIDIRECTIONAL GLENOHUMERAL INSTABILITY

- **Sulcus sign (Figure 4–17):**
  - The examiner pulls down on the patient's arm with one hand as they stabilize the scapula with the other. If an indentation develops between the acromion and the humeral head, the test is positive. This suggests increased laxity in the GH joint.

FIGURE 4–17 Sulcus sign.
Source: Courtesy of JFK Johnson Rehabilitation Institute, 2000.

Imaging

- X-ray films with AP, scapular-Y, and axillary lateral views
  - Axillary views assess for glenohumeral dislocations.
  - Other views:
    - West Point lateral axillary: Bankart lesions
    - Stryker notch view: Hill–Sachs lesions

Treatment

TRAUMATIC UNIDIRECTIONAL GLENOHUMERAL INSTABILITY (TUBS)

- *Conservative:*
  - Sling immobilization: Length of time is variable.
  - Rehabilitation: ROM and strengthening of the shoulder–girdle complex should follow the brief stage of immobilization.
    - Passive range of motion (PROM) with Codman's pendulum exercises
    - Isometric exercises early in the recovery course
- *Surgical:*
  - Muscle strengthening alone rarely prevents recurrent dislocations if there is sufficient capsular laxity. Surgery should be considered if rehabilitation fails in active individuals.
  - After a third dislocation, the risk for another approaches 100%. Surgery may then be considered. In athletes or active individuals, surgery may be considered earlier, particularly with a history of shoulder dislocation and instability associated with a labral tear.

POSTERIOR GLENOHUMERAL INSTABILITY

- Rehabilitation generally is adequate for the majority of these patients.
- *Conservative:*
  - Immobilize in a neutral position for roughly 3 weeks.
  - Strengthening the posterior shoulder–scapula musculature is imperative (infraspinatus, posterior deltoid, teres minor, trapezius, serratus anterior).
    - This phase may last up to 6 months.
- *Surgical:*
  - In the event of a failed rehabilitation program, a posterior capsulorrhaphy is the surgical procedure of choice for recurrent posterior shoulder dislocations of traumatic origin.

MULTIDIRECTIONAL GLENOHUMERAL INSTABILITY (AMBRI)

- >80% of the patients obtain excellent results with rehabilitation.
- Educating patients to avoid voluntarily dislocating the shoulder and to avoid positions of known instability should be part of the treatment program.
- Surgical treatment may be an option only when conservative measures fail. At that time, an inferior capsular shift may be indicated.

GLENOID LABRUM TEARS

General

- The labrum encircles the periphery of the glenoid fossa. Tendons (rotator cuff and biceps) insert on the labrum. As a result, any tear or instability of the labrum may be accompanied by rotator cuff or biceps tendon pathology.
- Repetitive overhead sports (baseball, volleyball) or traumas are causative factors.
- Tears may occur through the anterior, posterior, or superior aspect of the labrum.
- SLAP lesion: **S**uperior glenoid **L**abral tear in the **A**nterior-to-**P**osterior direction

Clinical Features

- Signs and symptoms are similar to that of shoulder instability (clicking, locking, pain).

PROVOCATIVE TESTS

- **Load-and-shift test:**
  - The examiner grasps the humeral head and pushes it into the glenoid while applying an anterior and posterior force. A positive test indicates labrum instability and is displayed by excess translation.
- 🕮 **O'Brien's test (Figure 4–18)**
  - Used to detect SLAP lesions
  - It is performed in two parts: With the arm internally rotated, forward flexed, and adducted about 15 degrees, the examiner applies a downward force to the patient's pronated arm initially. Then the examiner applies a downward force to the patient's supinated arm.
  - A positive test results in deep shoulder pain that improves when the downward force is applied with hand in supination.

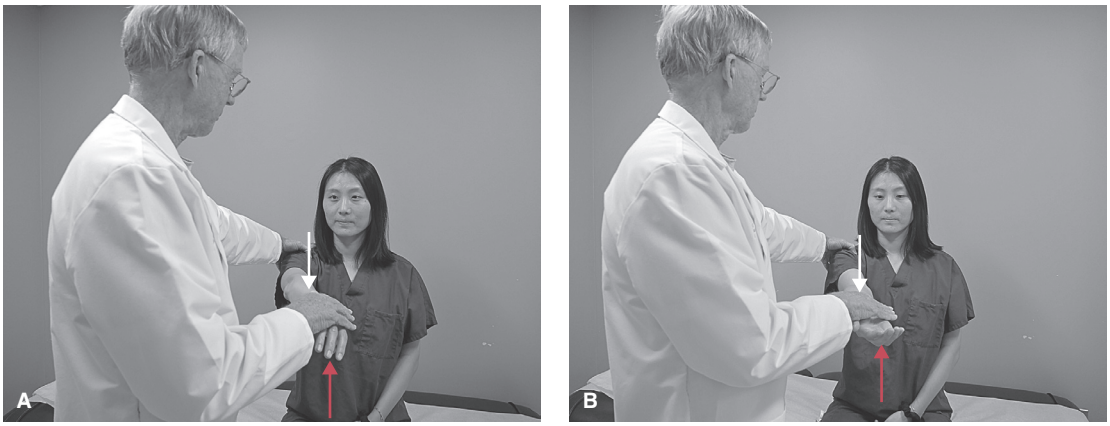

FIGURE 4–18 O'Brien's test. (A) With the arm internally rotated, forward flexed, and adducted about 15 degrees, the examiner applies a downward force to the patient's pronated arm initially (White arrow). (B) Then the examiner applies a downward force to the patient's supinated arm (White arrow). Red arrows indicate direction of resistance applied by the patient.

- **Anterior Slide Test (Figure 4–19):**
  - Used to detect SLAP tears
  - The patient is placed with their ipsilateral hand placed on the hips with the thumb aimed posteriorly.
  - The examiner places one hand on top of the patient's shoulder so that the last segment of the index finger extends over the acromion. Then the examiner applies force at the ipsilateral elbow in a forward and slightly upward motion, while the patient resists the movement.
  - The test is considered positive if the patient has pain in the anterior aspect of the shoulder or if there is an audible clicking/popping sensation.

FIGURE 4–19 Anterior slide test. The patient is placed with their ipsilateral hand placed on the hips with the thumb aimed posteriorly. The examiner places one hand on top of the patient's shoulder so that the last segment of the index finger extends over the acromion. Then the examiner applies force at the ipsilateral elbow in a forward and slightly upward motion (black arrow), while the patient resists the movement (white arrow).

Imaging and Treatment

- The same as GHJ instability

SHOULDER IMPINGEMENT SYNDROME AND ROTATOR CUFF TEAR

General

- **Impingement syndrome (Figure 4–20):**
  - Likely the most common cause of shoulder pain
  - A narrowing of the subacromial space causing compression and inflammation of the subacromial bursa, biceps tendon, and rotator cuff
  - Impingement of the rotator cuff tendon(s), **most commonly the supraspinatus**, under the acromion and the greater tuberosity occurs with arm abduction and internal rotation.
  - Impingement syndrome often leads to chronic tendinopathy, which can progress to a rotator cuff tear (partial or complete).

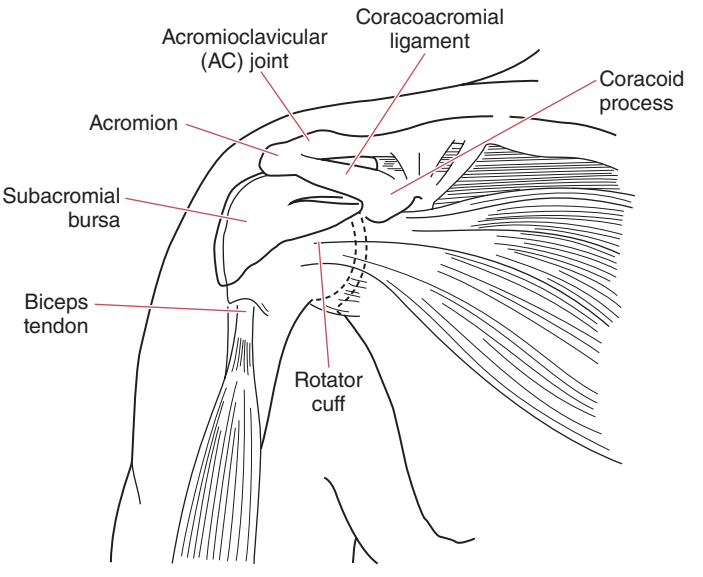

FIGURE 4–20 Anatomy of the shoulder (anterior view).

- **Stages of subacromial impingement syndrome (Neer, 1972):**
  - Stage 1: Edema or hemorrhage—reversible (age <25)
  - Stage 2: Fibrosis and tendonitis (ages 25–40)
  - Stage 3: AC spur and rotator cuff tear (age >40)

Rotator Cuff Tears

- The rotator cuff is composed of four muscles (S.I.T.S.; **Figure 4–21**):
  1. **S**upraspinatus
  2. **I**nfraspinatus
  3. **T**eres minor
  4. **S**ubscapularis
- These muscles form a cover around the head of the humerus and function to rotate the arm and stabilize the humeral head against the glenoid.
- 🕮 Rotator cuff tears occur primarily in the *supraspinatus tendon*, which is weakened as a result of many factors, including injury and chronic impingement. Poor blood supply to the tendon also makes it prone to injury, especially at the critical zone of hypovascularity about 1 cm from the insertion site.
- May be as a result of direct trauma or as an end result from chronic impingement. This injury rarely affects people younger than 40 years old.

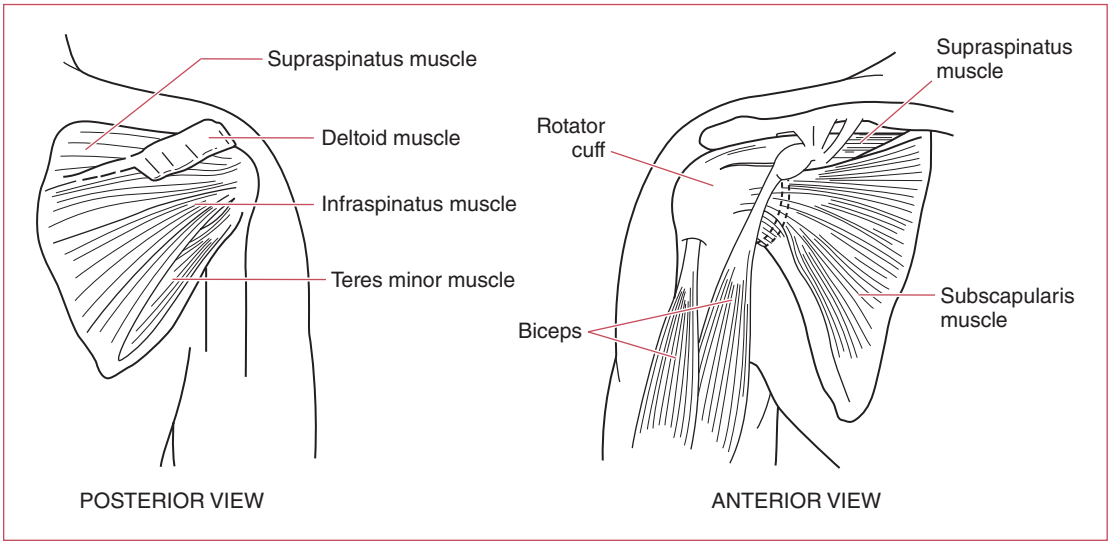

FIGURE 4–21 Rotator cuff muscles: Posterior view (left); anterior view (right).

ACROMION MORPHOLOGY AND ASSOCIATION TO ROTATOR CUFF TEARS

- Acromion morphology types **(Figure 4–22)**:
  - Type I → Flat
  - Type II → Curved
  - Type III → Hooked (Brown & Neumann, 1999)

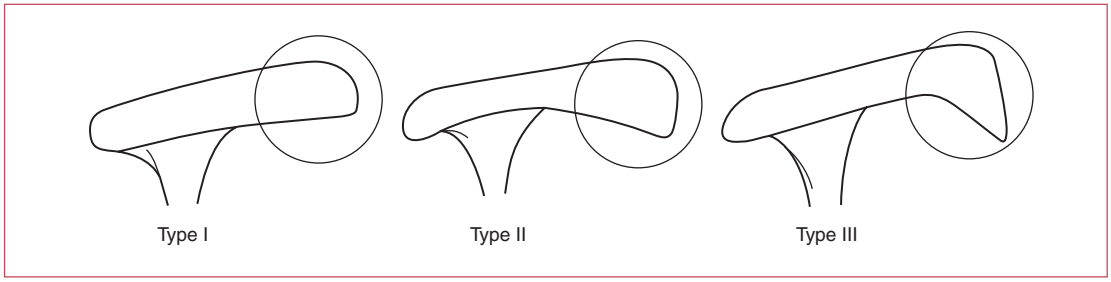

FIGURE 4–22 Three types of acromion morphology.

- The anatomic shape of the patient's acromion has been linked with occurrence rates of rotator cuff tears.
- Patients with a curved or hooked acromion have a higher risk of rotator cuff tears.

Clinical Features

- 🕮 Pain during ROM specifically in repetitive overhead activities, such as:
  - Throwing a baseball
  - Swimming
    - Phases of the swim stroke include the catch, propulsive pull and push, and recovery phases.
    - Occurs at the "catch" phase of the overhead swimming stroke
    - Mechanism: Flexion, abduction, internal rotation
    - More common strokes: Freestyle, backstroke, and butterfly
    - Less common stroke: Breaststroke
- The supraspinatus tendon is commonly affected secondary to its location under the acromion.
  - Patients may feel crepitus, clicking, or catching on overhead activities.
  - Pain may be referred anywhere along the shoulder girdle.
  - Pain and weakness felt in forward flexion, abduction, and internal rotation indicating impingement (Hawkins' sign)
  - Inability to initiate abduction may indicate a significant rotator cuff tear.
  - Pain may be nocturnal, as patients often report having difficulty sleeping on the affected side.
  - Tenderness observed over the greater tuberosity or inferior to the acromion on palpation
  - Atrophy of the involved muscle may occur, resulting in a gross deformity at the respective area, usually seen in large, chronic tears.

PROVOCATIVE TESTS

- **Impingement tests:**
  - **Neer's impingement sign (Figure 4–23):**
    - Stabilize the scapula and passively forward flex the arm >90 degrees.
    - Positive test with pain resulting from the supraspinatus tendon being compressed between the acromion and greater tuberosity of the humerus
  - **Hawkins' impingement sign (Figure 4–24):**
    - Stabilize the scapula and passively forward flex (to 90 degrees) the internally rotated arm.
    - Positive test with pain from the supraspinatus tendon being compressed between the coracoacromial ligament and greater tuberosity of the humerus
  - **Painful arc sign:**
    - Abducting the arm, with pain occurring roughly between 60 and 120 degrees
- **Rotator cuff tests:**
  - **Empty can (supraspinatus) test**
    - Pain and weakness with arm flexion abduction and internal rotation (thumb pointed down)
    - With abduction the humerus will naturally externally rotate. In assessing the integrity of the supraspinatus, the patient should internally rotate the humerus, forcing the greater tuberosity under the acromion. In this position, the maximum amount of abduction is to 120 degrees.
  - **Drop arm test:**
    - The arm is passively abducted to 90 degrees and internally rotated.
    - A positive test is noted when the patient is unable to maintain the arm in abduction or has difficulty lowering the abducted arm in a gradual and smooth fashion.
    - Initially the deltoid assists in abduction but fails quickly.
    - This indicates a complete tear of the cuff.

FIGURE 4–23 Neer's impingement sign. *Source:* Courtesy of JFK Johnson Rehabilitation Institute, 2000.

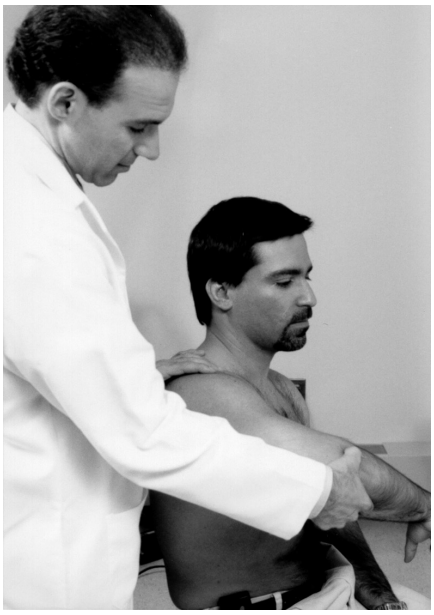

FIGURE 4–24 Hawkins' impingement sign. *Source:* Courtesy of JFK Johnson Rehabilitation Institute, 2000.

Imaging

- Plain films (AP):
  - Impingement: Typically normal. Can see cystic changes in the greater tuberosity chronically.
  - Chronic rotator cuff tear
    - Superior migration of the proximal humerus
    - Flattening of the greater tuberosity
    - Subacromial sclerosis
- Supraspinatus outlet view (30 degrees caudal tilt for a transcapular "Y" view; **Figure 4–25**):
  - Assess acromion morphology, evaluate for possible spur.
- 🕮 MRI is the gold standard to evaluate the integrity of the rotator cuff.
  - Full thickness tears and partial tears can be delineated.
  - Gadolinium may be added to evaluate the labrum.
- Arthrogram:
  - 🕮 Beneficial in assessing full thickness tears but unable to delineate the size of the tear or partial tears.
  - An MRI arthrogram may be indicated to evaluate patients suspected of having a labral tear that may not be seen on a routine shoulder MRI.
  - Should not be used in patients who have gadolinium contrast dye allergy.
- 🕮 Ultrasound (US):
  - Full thickness tears may be demonstrated by nonvisualization of the rotator cuff, indicating discontinuity of the cuff and interposition of the subacromial bursa or deltoid into the vacant tendon.
  - Thickened, heterogeneous appearing tendon, cortical irregularity, or defect in the cuff tendon may indicate partial tear or tendinosis.
  - Quality of the assessment is operator-dependent.

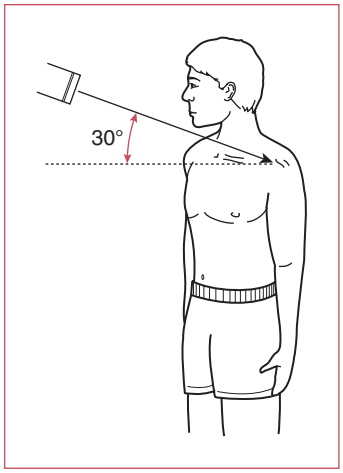

FIGURE 4–25 Radiography of the rotator cuff: 30-degree angled view.

Treatment

IMPINGEMENT, CHRONIC PARTIAL, AND FULL TEARS

- *Conservative* (*Rehabilitation*):
  - **Acute phase (up to 4 weeks)**
    - Relative rest: Avoid any activity that aggravates the symptoms.
    - Reduce pain and inflammation.
    - Modalities: US, iontophoresis
    - Reestablish nonpainful and scapulohumeral ROM.
    - Retard muscle atrophy of the entire upper extremity.
  - **Recovery phase (months)**
    - Improve upper extremity ROM and proprioception.
    - Full pain-free ROM
    - Improve rotator cuff (supraspinatus) and scapular stabilizers (rhomboids, levator scapulae, trapezius, serratus anterior).
    - Assess single planes of motion in activity-related exercises.
  - 🕮 **Functional phase**
    - Continue strengthening, increasing power and endurance (plyometrics).
    - Perform activity-specific training.
    - Rehabilitation in swimmers focuses on strengthening the rotator cuff muscles and scapular stabilizers, including serratus anterior and lower trapezius.
    - Corticosteroid injection: Only up to three injections yearly (may weaken the collagen tissue, leading to more microtrauma)
- *Surgical:*
  - **Indications**
    - Partial or full thickness tears that fail conservative treatment
    - Reduction or elimination of impingement pain is the primary indication for surgical repair in chronic rotator cuff tears. The patient should be made aware that restoration of abduction is less predictable than relief of pain.
  - *Partial tears* (*<40% thickness*)
    - Procedure: Partial anterior acromioplasty and coracoacromial ligament lysis
  - *Partial tears* (*>40% thickness*)
    - Repair rotator cuff tendon.
  - *Acute rotator cuff tears* (*i.e., athletes/trauma*)
    - Statistics show that surgical repair of an acute tear within the first 3 weeks results in significantly better overall function than later reconstruction.

DEGENERATIVE JOINT DISEASE OF THE SHOULDER (FIGURE 4–26; OSTEOARTHRITIS OF THE SHOULDER)

General

- Destruction of the articular cartilage and narrowing of the joint space
- Arthritis may occur at the GH or AC joint.
- It is also seen in posttraumatic lesions, chronic rotator cuff pathology, Lyme disease, and more.

Clinical Features

- Limitations and pain on active and passive ROM, which lead to impairment of activities of daily living (ADLs)

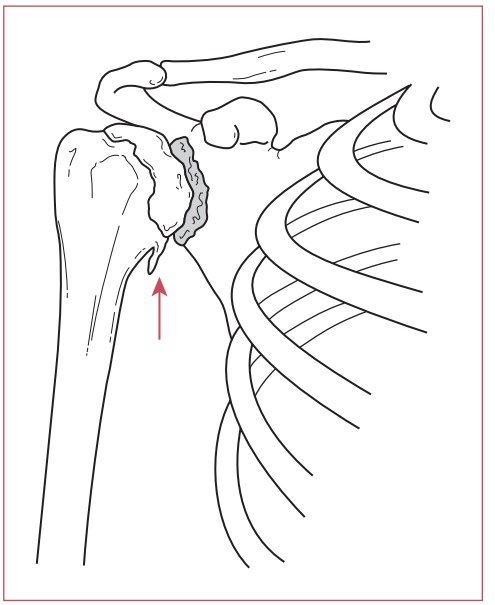

FIGURE 4–26 Degenerative joint disease of the GH joint.
GH, glenohumeral.

- Pain more common in internal rotation of the shoulder but may also be seen with abduction
- Manual muscle testing may or may not be affected depending on the severity of the disease.
- Pain may be nocturnal and relieved by rest.
- Tenderness felt on palpation on the anterior and posterior aspects of the shoulder

Imaging

- X-ray AP view: Internal and external rotation and 40 degrees of obliquity
- Axillary view
- Changes seen on x-ray include:
  - Irregular joint surfaces
  - Joint space narrowing (cartilage destruction)
  - Subacromial sclerosis
  - Osteophyte changes
  - Flattened glenoid
  - Cystic changes in the humeral head

Treatment

- *Conservative:*
  - Goal is to decrease pain and inflammation.
  - NSAID, corticosteroid injection
  - Rehabilitation
  - ROM and rotator cuff strengthening
- *Surgical:*
  - **Total shoulder arthroplasty (TSA)**
    - Indications
      - Pain
      - Avascular necrosis (AVN)
      - Neoplasm
    - Goals: Relieve pain, protect joint, and restore function.
  - **Stage 1: 0 to 6 weeks**
    - Precautions status post-TSA
      - Avoid active abductions and extension >0 degrees.
      - Sling immobilization
      - No external rotation >15 degrees
      - No active ROM, non-weight bearing
    - Treatment: Gentle PROM (Codman's exercises), gentle active range of motion (AROM; wall-walking), isometric exercises (progressing)
  - **Stage 2: 6 to 12 weeks**
    - Precautions: Discontinue sling, start light weights.
    - Treatment: Isotonics, active-assist ROM (AAROM), AROM
  - **Stage 3: >12 weeks**
    - Precautions: Previous ROM precautions cancelled
    - Treatment: Start progressive resistive exercises, active ranging, and stretching.
  - **Reverse TSA**
    - Used for rotator cuff arthropathy associated with chronic rotator cuff deficiency
  - **Shoulder arthrodesis**
    - Surgical resection and fusion of the GHJ
    - Typical patient is a young heavy laborer with repetitive trauma to the shoulder.
    - Indications
      - Severe shoulder pain secondary to osteoarthritis (OA)
      - Mechanical loosening of a shoulder arthroplasty
      - Joint infection

- 🕮 Fusion position
  - 50-degree abduction
  - 30-degree forward flexion
  - 50-degree internal rotation

CALCIFIC TENDONITIS OF THE SUPRASPINATUS TENDON (ROTATOR CUFF CALCIFIC TENDINOPATHY)

General

- Calcium deposits, most commonly involving the supraspinatus tendon
- Size of the deposit has no correlation to symptoms.

Clinical Features

- Sharp pain in the shoulder with ROM, particularly with shoulder abduction and overhead activities

Imaging

- AP x-ray of the shoulder will show calcium deposits, usually at the tendon insertion site.

Treatment

- Symptoms can improve with subacromial injection and physical therapy.
- US-guided percutaneous needling, aspiration, and saline lavage of the calcific lesion have been performed with successful results.
- Surgical treatment is rare and reserved for patients with severe pain and inability to perform ADLs who have failed more conservative treatments.

ADHESIVE CAPSULITIS (FROZEN SHOULDER; FIGURE 4–27)

General

- Painful shoulder with restricted GH motion
- Contracture of the shoulder joint
- Unclear etiology: May be autoimmune or trauma, but prevailing hypothesis suggests that within the joint capsule and synovial fluid, inflammation first occurs and is followed by reactive fibrosis and adhesions in the synovial lining.
- More common in women over the age of 40 years
- Associated with a variety of conditions:
  - Intracranial lesions: Cerebral vascular accidents (CVAs), hemorrhage, and brain tumor
  - Clinical depression
  - Shoulder–hand syndrome
  - Parkinson's disease
  - Iatrogenic disorders (prolonged immobilization)
  - Cervical disc disease
  - Insulin-dependent diabetes mellitus (DM)
  - Hypothyroidism

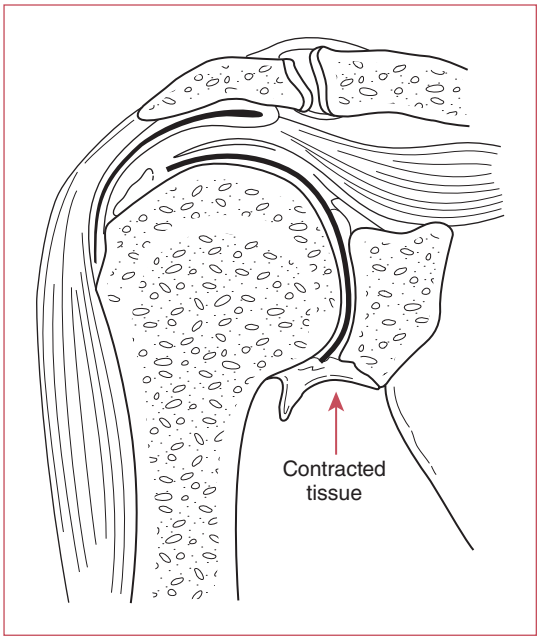

FIGURE 4–27 Glenohumeral joint in adhesive capsulitis. Note thickened and contracted capsular tissue.

STAGES
- Painful stage: Progressive vague pain lasting roughly 8 months
- Stiffening stage: Decreasing ROM lasting roughly 8 months
- Thawing stage: Increasing ROM with decrease of shoulder pain

PATHOLOGY
- Synovial tissue of the capsule and bursa become adherent.

Clinical Features
- Pain, with significant reduction in both AROM and PROM
- External rotation and abduction ROM typically lost first. Shoulder flexion, adduction, and extension are subsequently lost.

Imaging
- Plain films (AP view)—typically normal but indicated to rule out underlying tumor or calcium deposit. Indicated in patients whose pain and motion do not improve after 3 months of treatment.
- Osteopenia may be seen; otherwise, normal.
- Shoulder MRI demonstrates thickened GHJ capsule and synovium.
- Shoulder arthrogram will demonstrate a decreased volume in the joint, which can be realized by the small amount of contrast dye (<5 mL) that can be injected.

Treatment
- *Conservative:*
  - Restoring PROM and AROM
  - Decreasing pain
  - Corticosteroid injection: Subacromial and GH can decrease pain to maximize therapy. Variable efficacy.
  - Home program: Stretches in all ranges of motion
  - Modalities: US and electrical stimulation
- *Surgical:*
  - Manipulation under anesthesia may be indicated if there is no substantial progress after 12 weeks of conservative treatment.
  - Arthroscopic lysis of adhesions—usually reserved for patients with insulin-dependent diabetes mellitus (IDDM) who do not respond to manipulation.

BICEPS TENDONITIS AND RUPTURE

General

BICEPS TENDONITIS (FIGURE 4–28)
- Origin of the long head of the biceps tendon is the supraglenoid tuberosity.
- Origin of the short head of the biceps tendon is the apex of the coracoid process.
- Typically inflammation of the long head of the biceps tendon occurs at the bicipital groove of the humeral head.
- This tendon may be impinged between the head of the humerus, acromion, and CC ligaments with elevation and internal rotation of the arm.

🕮 Biceps Tendon Rupture (Figure 4–29)
- Most common site of rupture is at the proximal end of the long head of the biceps tendon.
- "Popeye sign" noted with retracted biceps muscle

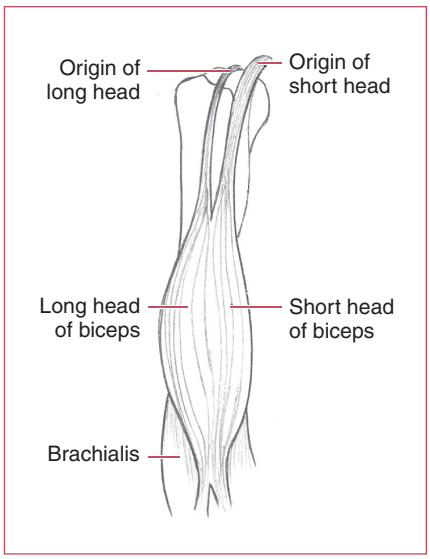

FIGURE 4–28 Anterior muscles of the right arm.

- Distal rupture is rare, typically occurring in people performing significant physical activities (e.g., bodybuilders, heavy manual workers).
- Seen in adults >40 years old with a chronic history of impingement syndrome
- Also associated with rotator cuff tears in the elderly

Clinical Features

- Point tenderness in the bicipital groove **(Figure 4–30)**
- Positive impingement signs if associated with shoulder impingement syndrome
- Sharp pain, audible snap, ecchymosis, and visible bulge ("Popeye muscle") in the upper arm with tendon rupture

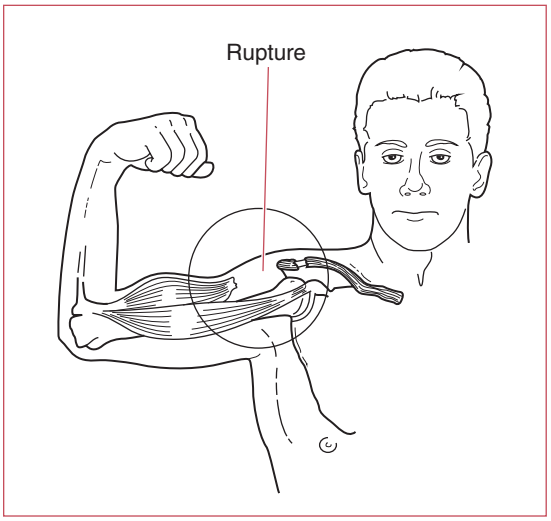

FIGURE 4–29 Rupture of the proximal biceps tendon (rupture is better appreciated on attempted contraction).

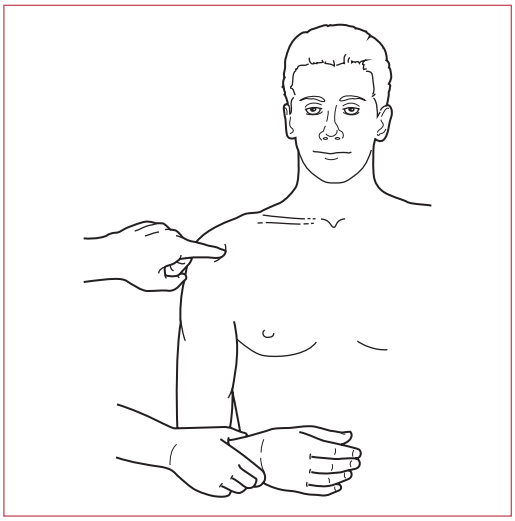

FIGURE 4–30 Point tenderness of biceps tendon in bicipital groove.

PROVOCATIVE TESTS

- *Biceps tendonitis:*
  - **Yergason's test (Figure 4–31)** determines the stability of the long head of the biceps tendon in the bicipital groove.
    - Pain at the anterior shoulder with flexion of the elbow to 90 degrees and supination of the wrist against resistance

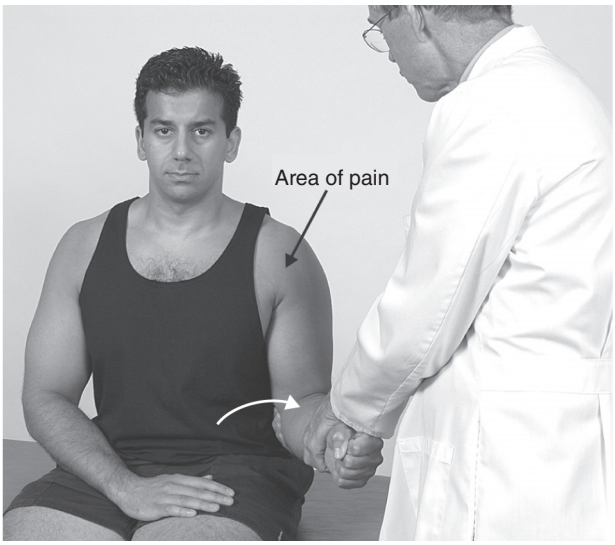

FIGURE 4–31 Yergason's test. Pain may be elicited in the anterior shoulder when the patient supinates the wrist/forearm against resistance. *Source:* Courtesy of JFK Johnson Rehabilitation Institute, 2000.

  - **Speed's test:**
    - Pain at the anterior shoulder with flexion of the shoulder, elbow extended, and supinated against resistance
- *Biceps rupture*:
  - **Ludington's test:**
    - With the patient's hands resting on top of their head (finger interlocked), the patient is asked to contract and relax the biceps muscles on each side.
    - With palpation of the long head biceps groove during biceps contraction, contraction of the biceps tendon will be absent on the affected side, while it can be felt on the unaffected side.

Imaging

- Nonspecific findings on plain radiographs
- MRI may show tendinopathy or biceps tendon rupture.
- Diagnostic US imaging can provide quick, in-office evaluation of an acute biceps tendon rupture.

Treatment

- Tendonitis:
  - Conservative treatment is appropriate for most patients.
  - ROM and strengthening as tolerated
  - Modalities
  - Injection into the tendon sheath (controversial due to the potential for tendon rupture)
- Rupture:
  - Tendon reattachment is not indicated in most patients.
  - Biceps tenodesis can be considered in younger, active individuals who require heavy lifting.
  - Some patients may request reattachment of biceps tendon for cosmetic reasons.

DELTOID STRAIN AND AVULSION

General

- The deltoid muscle arises from the anterior clavicle, the acromion, and the spine of the scapula.
- Primarily innervated by the axillary nerve
- Involved with flexion, extension, and abduction of the GHJ
- Complete rupture of the deltoid is rare.
  - Deltoid rupture is most commonly associated with surgical intervention but may also occur with crush injuries or severe direct blows.
- Usually strains and contusions occur with direct blow to the upper arm when it is in abduction and forward flexion.
- Anterior deltoid can be injured during the acceleration phase of throwing.
- Posterior deltoid can be injured during the deceleration phase of throwing.

Clinical Features

- May injure the origin of the deltoid with Grade V AC joint separations
- Swelling, local tenderness, and limited shoulder motion may occur with strains without rupture.
- With an acute rupture, there may be swelling, deformity, ecchymosis, palpable defect, and weakness.

Imaging

- Plain radiographs will likely be normal unless there is significant injury (i.e., rupture) with associated injury (e.g., shoulder dislocation).
- MRI of the shoulder can better assess soft tissue pathology in suspected cases of deltoid rupture.

Treatment

- For strains and contusions, ice and immobilize acutely. Then perform stretching and progressive strengthening exercises.
- For complete rupture or avulsion, treatment is surgical reattachment.

SCAPULAR WINGING (FIGURE 4–32)

(Also see Chapter 5, Electrodiagnostic Medicine and Clinical Neuromuscular Physiology, Table 5–31).

General

TYPES

- **Medial scapular winging:**
  - Results from serratus anterior weakness
  - Often the result of a long thoracic nerve palsy
  - Bench pressing very heavy weights or wearing heavy pack straps can also impinge the nerve.
  - Scapula is elevated and retracted.
- **Lateral scapular winging:**
  - Results from trapezius muscle weakness
  - Can be due to spinal accessory nerve lesions.
  - Nerve injury occurs in the posterior triangle of the neck.
  - Scapula is depressed and protracted.

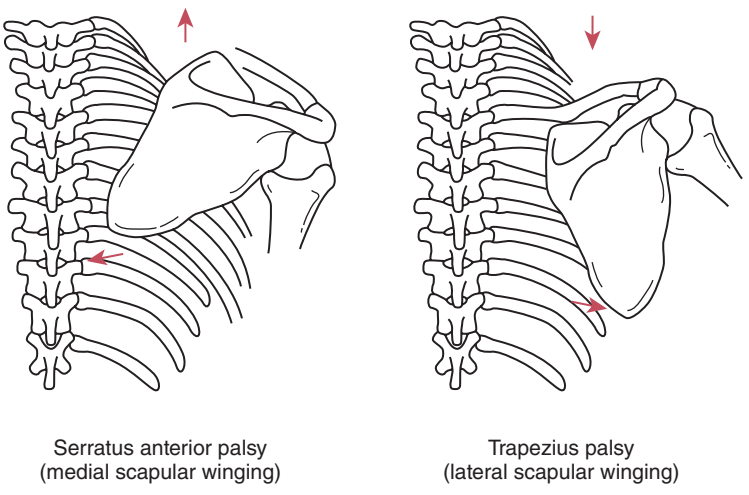

FIGURE 4–32 Scapular winging patterns.

Clinical Features

- **Medial scapular winging:**
  - Winging of the medial border of the scapula away from the ribs
  - More evident when the patient forward flexes the arms or does a wall push-up
- **Lateral scapular winging:**
  - Rotary lateral winging of the scapula around the thorax
  - Upper trapezius muscle fibers can be tested by resisted shrug.
  - Middle and lower trapezius fibers can be tested by prone rowing exercise.
- Electrodiagnostic studies should be considered to diagnose nerve injury and prognosis.

Imaging

- Often not directly helpful
- Type of winging will determine specific imaging workup.

Treatment

- Scapular stabilization rehabilitation

SCAPULAR FRACTURES (FIGURE 4–33)

General

- Scapular fractures commonly occur in association with other serious injuries. The diagnosis often is easily missed on the initial exam.
- Mechanism typically is a direct blow to the shoulder usually after a significant, high-velocity trauma (e.g., motor vehicle accidents [MVA], motorcycle accident).
- Associated with other significant injuries such as rib fractures, pulmonary contusions, and pneumothorax/hemothorax
- Fracture sites: Glenoid, glenoid rim, coracoid, scapular neck and body, acromion

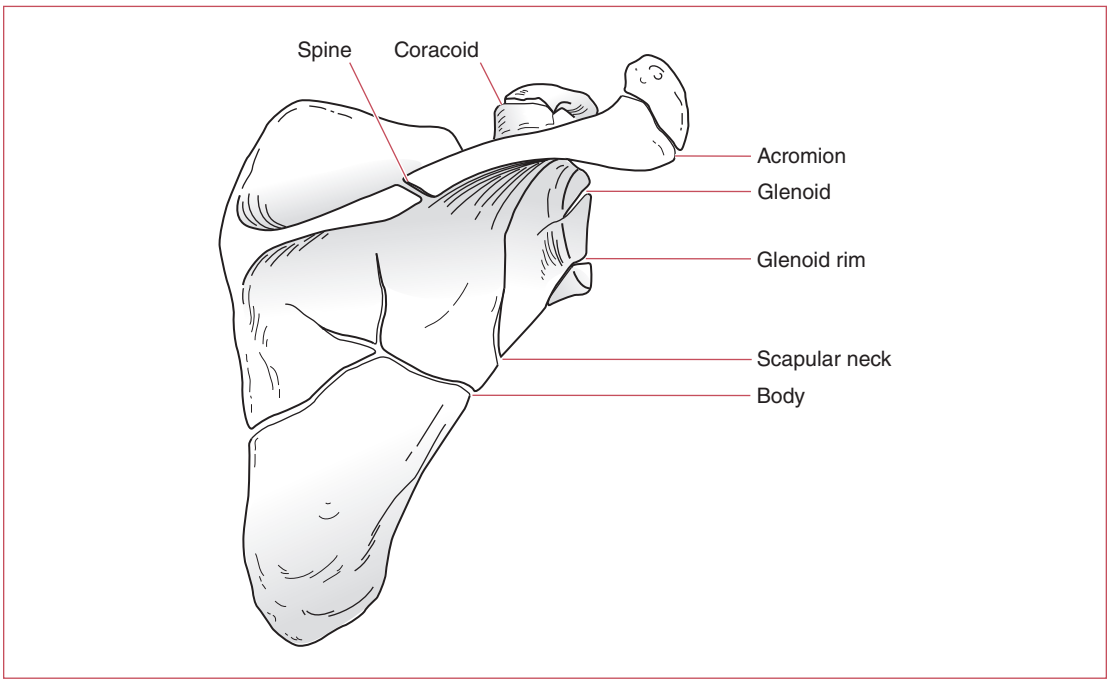

FIGURE 4–33 Scapular fracture patterns.

Clinical Features

- Tenderness over the scapular and acromial region

Imaging

- Plain films: AP, lateral scapular-Y, and axillary views
- CT scan

Treatment

- Closed treatment is adequate for nondisplaced fragments.
- Arm sling followed by early ROM exercises as tolerated, usually within 1 to 2 weeks after injury
- ORIF: Large displaced fragments
- Note: Patients with isolated scapular body fractures should be considered for hospital admission due to the risk of pulmonary contusion.

CLAVICULAR FRACTURES

General

- Classification is based on fracture location
- Fracture located at medial, middle (most common), or distal third of the clavicle

Clinical Features

- Pain, swelling, ecchymosis in the shoulder/clavicular region, typically after trauma such as a fall or direct impact. May or may not have an obvious deformity.
- AC joint and sternoclavicular (SC) joints should also be assessed, as they may also be injured.

Imaging

- AP plain films of the clavicle with inclusion of AC and SC joints. Chest x-ray to evaluate for superimposed pneumothorax complication.

Treatment

- Most clavicular fractures can be treated conservatively.
  - Closed reduction and immobilization with a simple sling or figure-8 sling
  - Immobilization may range from 3 to 6 weeks depending on the age.
  - Progressive ROM may be initiated after 3 weeks of immobilization.
- Surgery indicated for open clavicle fractures, grossly displaced fracture with skin tenting, and fractures with significant medialization of shoulder girdle
  - Displaced lateral clavicle fractures (>1 cm) at the AC joint are best treated surgically.

PROXIMAL HUMERAL FRACTURES

General

- Occur primarily in older osteoporotic patients after a low-energy fall, or in young patients who experience a high-energy trauma
- Account for approximately 5% of all fractures
- Classification is based on the Four-Part Classification.
- This classification involves displacement of fractures in four different parts of the humerus in relation to each other (Snider, 1997). These areas are:
  - Greater tuberosity
  - Lesser tuberosity
  - Humeral head
  - Humeral shaft
- One of these parts must be angulated by 45 degrees or displaced at least 1 cm to be considered displaced.

Four-Part Classification (Figure 4–34)

- One-part humeral fracture: Nondisplaced, impacted fractures. All parts still in alignment.
- Two-part humeral fracture: One fragment is displaced with respect to the other three.
- Three-part humeral fracture: Two fragments are displaced.
- Four-part humeral fracture: All fragments are displaced.
- Common locations for fractures include:
  - Greater tuberosity
  - Lesser tuberosity
  - 🕮 Surgical neck (most common)
  - Anatomic neck

Clinical Features

- Mechanism: Most commonly from a fall on an outstretched hand, usually from a standing height. Thus, most fractures are from an indirect blow; however, direct impact can also cause fractures.

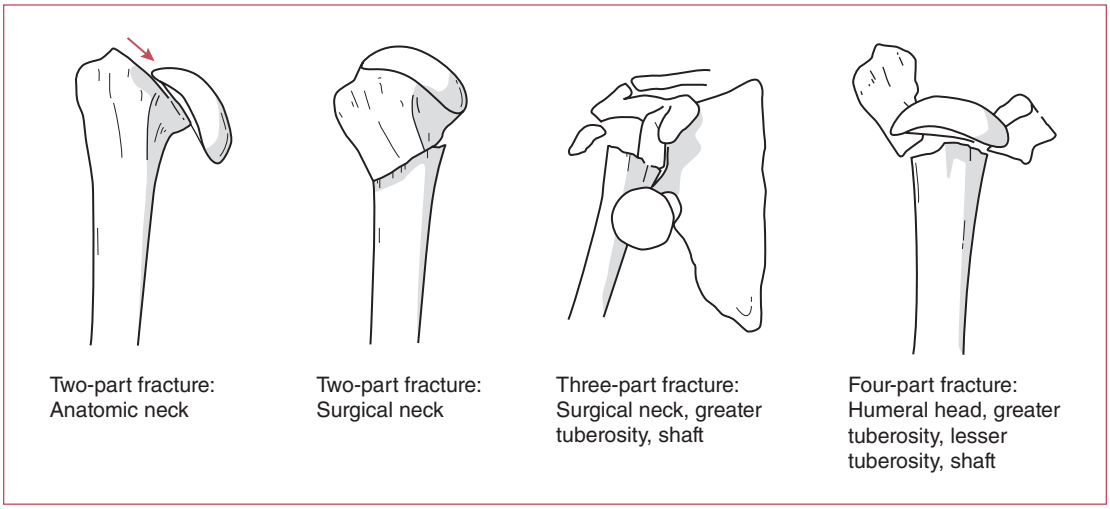

FIGURE 4–34 Displaced proximal humerus fracture patterns (Neer classification).

- Typically occurs in elderly women with osteoporosis after a fall.
- Pain, swelling, and ecchymosis in the upper arm, exacerbated with the slightest motion
- 🕮 In fracture at the surgical neck, the supraspinatus is the principal abductor (i.e., supraspinatus causes abduction of the proximal fragment of the humerus).
- Loss of sensation is seen if there is neurologic involvement.
- Diminished radial pulse if the fracture compromises the vascular supply

Imaging

- X-ray (trauma series): AP view, scapular-Y view, axillary view, apical oblique view, and West Point axillary view

Treatment

- One part (nondisplaced):
  - *Conservative:* Sling immobilization and early rehabilitation (6 weeks) – Early ROM: Codman's exercises and AROM as early as tolerated – AROM, pendulum exercises as early as tolerated.
- *Surgical:* ORIF:
  - Greater than one part (displaced >2 cm)

Complications

- Neurovascular:
  - 🕮 Brachial plexus injuries
  - 🕮 Axillary nerve injury can occur with surgical neck fractures.
  - Radial and ulnar nerves may be affected as well.
  - Median nerve is the least affected.
  - Axillary artery compromise may be evident depending on the site of injury.
- AVN of the humeral head may occur with anatomic neck fractures secondary to interruption of the humeral circumflex artery.

STRESS FRACTURES OF THE HUMERUS

General

- Stress fractures of the epiphyseal growth plate (epiphysiolysis) in the proximal humerus, also referred to as Little Leaguer's shoulder, occur through the proximal growth plate of the proximal humerus in skeletally immature overhead athletes, such as pitchers.

- Stress fractures of the humerus in adults may occur at the shaft.
- Repetitive torsional forces and opposing muscular contractions during throwing are the likely causes.

Clinical Features

- Insidious onset of shoulder pain aggravated by repetitive overhead throwing
- Focal tenderness over the stress fracture
- Discomfort with resistance to shoulder abduction and internal rotation
- Mild weakness may be possible.

Imaging

- Early plain films may be unremarkable.
- With chronic stress fractures, there may be cortical thickening along the mid-third of the medial cortex.
- In adolescent pitchers, widening of the lateral part of the physis with associated sclerosis or cystic changes may be seen on external rotation AP films.

Treatment

- Symptoms usually resolve with activity restriction of 8 weeks in adults and 12 weeks in adolescents.
- Continuing with precipitating factors may lead to spiral fracture of the humerus or premature closure of the physis.
- Return to gradual throwing when asymptomatic.

UPPER EXTREMITIES: THE ELBOW REGION

FUNCTIONAL ANATOMY

Elbow Joint Articulations

- Humeroulnar joint
- Humeroradial joint
- Proximal radioulnar joint

Elbow Range of Motion

- Elbow flexion: 135 degrees
- Elbow extension: 0 to 5 degrees
- Forearm supination: 90 degrees
- Forearm pronation: 90 degrees

Elbow Motion

- **Elbow flexion (Figure 4–35):**

ELBOW FLEXORS	SPINE INNERVATION	PERIPHERAL INNERVATION
Brachialis	C5, C6, C7	Musculocutaneous nerve from lateral cord
Biceps brachii	C5, C6	Musculocutaneous nerve from lateral cord
Brachioradialis	C5, C6, C7	Radial nerve from posterior cord
Pronator teres (PT)	C6, C7	Median nerve from lateral cord

- **Elbow extension (Figure 4–36):**

ELBOW EXTENSORS	SPINE INNERVATION	PERIPHERAL INNERVATION
Triceps	C6, C7, C8	Radial nerve from posterior cord
Anconeus	C7, C8, T1	Radial nerve from posterior cord

- **Forearm supination (Figure 4–37):**

FOREARM SUPINATORS	SPINE INNERVATION	PERIPHERAL INNERVATION
Supinator	C5, C6	Posterior interosseous nerve (PIN; radial nerve) from posterior cord
Biceps brachii	C5, C6	Musculocutaneous nerve from lateral cord

- **Forearm pronation (Figure 4–38):**

FOREARM PRONATORS	SPINE INNERVATION	PERIPHERAL INNERVATION
Pronator teres	C6, C7	Median nerve from lateral cord
Pronator quadratus	C7, C8, T1	Anterior interosseous nerve (AIN; median nerve)
Flexor carpi radialis	C6, C7	Median nerve from lateral cord

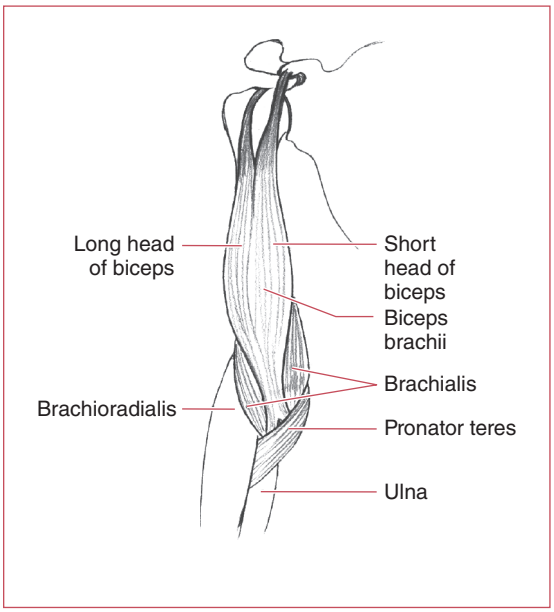

FIGURE 4–35 Elbow flexors (anterior view).

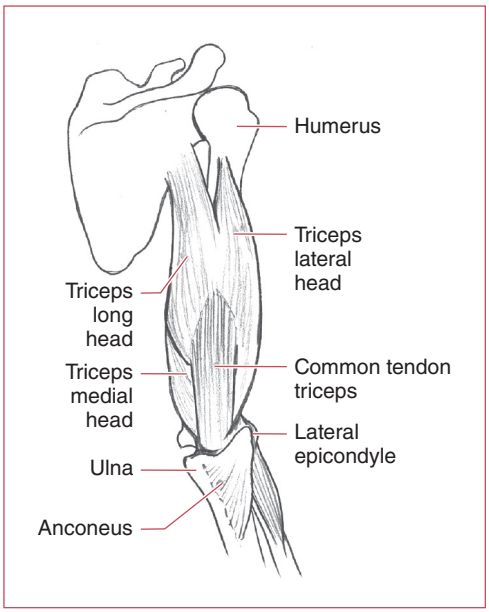

FIGURE 4–36 Elbow extensors (posterior view).

Elbow Ligaments (Figure 4–39)

- **Medial (ulnar) collateral ligament (MCL):**
  - Key stabilizer of the elbow joint (anterior band)
- **Lateral (radial) collateral ligament (LCL)**
- **Annular ligament:**
  - Holds the radial head in proper position

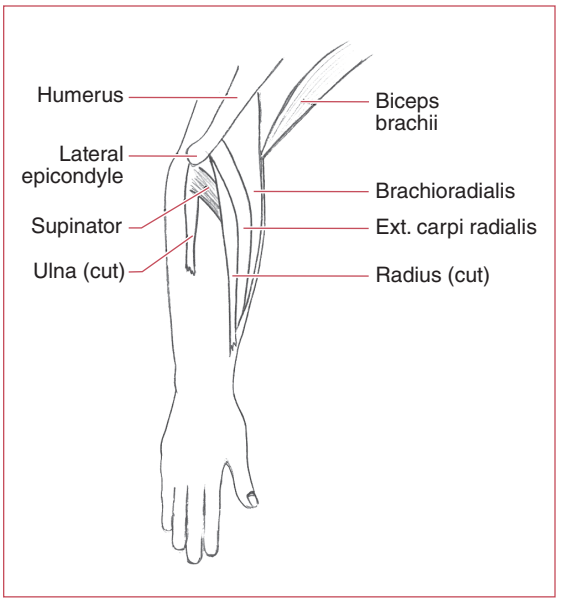

FIGURE 4–37 Forearm supinators (dorsal view).

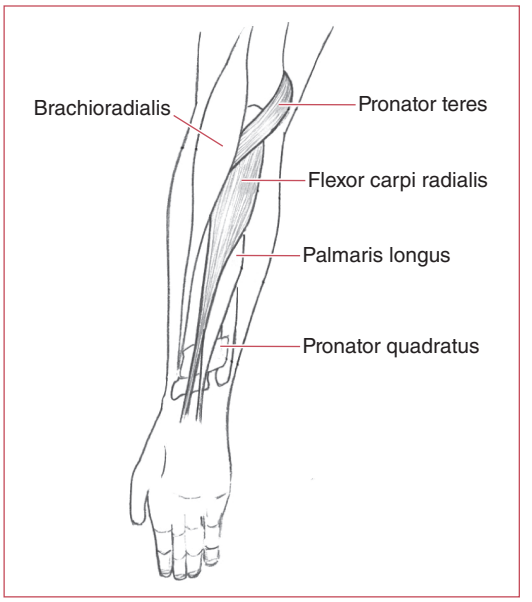

FIGURE 4–38 Forearm pronators (dorsal view).

Common Muscle Origins at the Elbow Joint

- **Medial epicondyle of the humerus:**
  - Flexor carpi radialis (FCR)
  - Flexor digitorum superficialis (FDS)
  - Flexor digitorum profundus (FDP)
  - Palmaris longus
  - Pronator teres (PT)
  - Flexor carpi ulnaris (FCU)
- **Lateral epicondyle of the humerus:**
  - Extensor carpi radialis longus (ECR-L)
  - Extensor carpi radialis brevis (ECR-B)
  - Extensor carpi ulnaris (ECU)
  - Extensor digitorum superficialis (EDS)
  - Supinator
  - Anconeus

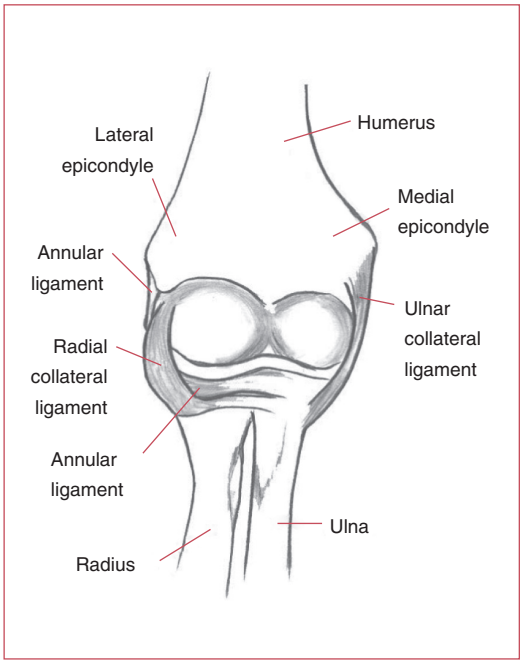

FIGURE 4–39 Elbow ligaments (anterior view of right elbow).

Mechanics of the Elbow: Carrying Angle

- The carrying angle is the anatomic valgus angulation between the upper arm and forearm when the arm is fully extended.
- It allows for the arm to clear the body when it is extended and supinated.
- Normal carrying angle (from anatomic position):
  - Males: 5 degrees of valgus
  - Females: 10 to 15 degrees of valgus
  - Angle >20 degrees is abnormal.

Elbow Arthrodesis

- Indications:
  - Arthritis
  - Failed surgical procedure

- 🕮 *Fusion position:*
  - Unilateral: Flexion—90 degrees
  - Bilateral: Flexion—110 degrees in one arm and 65 degrees for the other

ELBOW DISORDERS

MEDIAL EPICONDYLITIS

General

- Also known as golfer's elbow or pitcher's elbow

MECHANISM

- Caused by repetitive valgus stress to the elbow
- More commonly seen in athletes, especially in baseball pitchers and golfers. The throwing motion of a pitcher (especially in the late cocking and acceleration phase) and swinging motion (backswing and downward follow-through swing just prior to ball impact) of a golfer both place significant valgus stress on the elbow (Figure 4–40).
- Also occurs from the back and downward motion of a golf swing just prior to the impact of the ball

Biomechanics of Throwing a Baseball—Four Phases (Figure 4–40)

- Early cocking phase
- Late cocking phase
- Acceleration phase
- Follow-through

FIGURE 4–40 Throwing mechanics. (A) Early cocking phase. (B) Late cocking phase. (C) Acceleration phase. (D) Follow-through.
Source: Courtesy of JFK Johnson Rehabilitation Institute, 2000.

PATHOLOGY

- Inflammation of the common flexor tendon at the elbow
- Recurrent microtrauma can affect all medial elbow structures, which include the medial epicondyle, the medial epicondylar apophysis, and the ulnar collateral ligament (UCL) of the elbow, which may cause hypertrophy of the medial epicondyle.
- **Little Leaguer's Elbow (Medial epicondyle apophysitis of the elbow):**
  - Long-term repetitive valgus stress loading to the elbow in children, who have immature bones, can lead to medial epicondylitis and traction apophysitis of the medial epicondyle as a result of the recurrent microtrauma.
  - Hypertrophy of the medial epicondyle leading to microtearing and fragmentation of the medial epicondylar apophysis
  - May lead to osteochondritis dissecans of the capitellum

Clinical Features

- Tenderness just distal to the medial epicondyle over the common flexor tendon origin
- Pain may be reproduced with resisted wrist flexion and pronation.
- Ulnar neuropathy symptoms may occur secondary to valgus stretch of the nerve.

Imaging

- X-ray may show physeal widening and/or avulsion of the medial epicondyle.
- MRI can show edema in the medial epicondyle apophysis.

Treatment

- *Conservative:*
  - Short term: Rest, ice, NSAIDs, immobilization
  - Long term: Activity and modification of poor throwing mechanics extremely important
- *Surgical:*
  - Surgical pinning: Reserved for an unstable elbow joint

LATERAL EPICONDYLITIS

General

- Commonly known as tennis elbow

MECHANISM OF INJURY

- Activities that require repetitive wrist extension and/or forearm supination
- Common in racquet sports like tennis. Also seen in golfers.
- Overuse and poor mechanics lead to an overload of the wrist extensor tendons.
- Poor technique with racquet sports:
  - Improper technique for backhand swings
  - Inappropriate string tension
  - Inappropriate grip size

PATHOLOGY

- Microtearing of the ECR-B

Clinical Features

- Tenderness just distal to the lateral epicondyle at the extensor tendon origin
- Pain and weakness in grip strength

PROVOCATIVE TEST

- 🕮 **Cozen's test (Figure 4–41A):**
  - The examiner stabilizes the elbow with a thumb over the extensor tendon origin just distal to the lateral epicondyle. Pain in the lateral epicondyle is seen with the patient making a fist,

pronating the forearm, and radially deviating and extending the wrist against resistance by the examiner. (The test may be more sensitive when done in full extension at the elbow.)

- **Mill's test (Figure 4–41B):**
  - Passive extension of the elbow with forced flexion of the wrist with radial deviation may precipitate pain at the lateral epicondyle.
- **Maudsley's test (Figure 4–41C):**
  - While the patient is actively extending their third finger, the examiner applies resistance to it and palpates the lateral epicondyle, which can elicit pain in a positive test (DeLee et al., 2010).

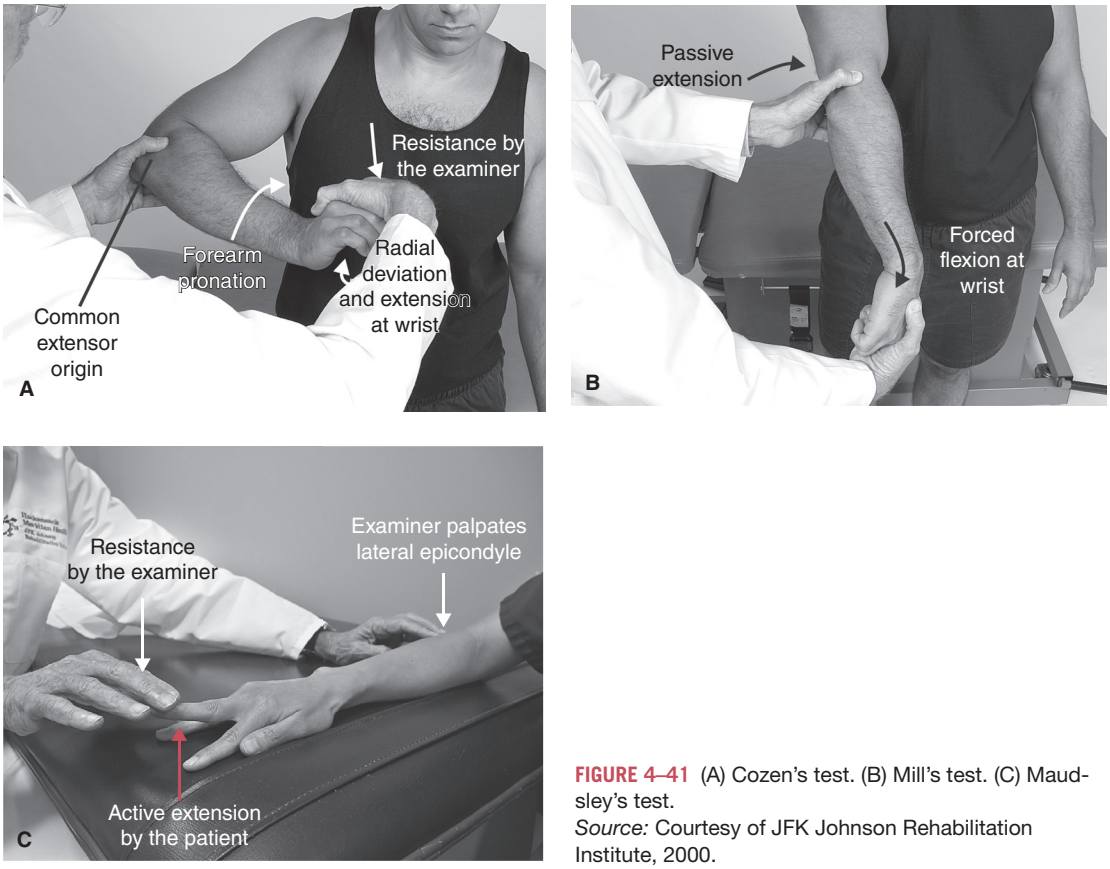

FIGURE 4–41 (A) Cozen's test. (B) Mill's test. (C) Maudsley's test.
Source: Courtesy of JFK Johnson Rehabilitation Institute, 2000.

Imaging

- Plain films of the elbow if arthritis and/or loose body fragments are suspected
- MRI to evaluate a tear in the common wrist extensor tendon, notably the ECR-B tendon

Treatment

- *Conservative:*
  - Relative rest, ice, NSAIDs for 10 to 14 days
  - Physical therapy (stretching, strengthening, modalities)
  - Splinting, bands
  - Corticosteroid injection controversial in its efficacy
  - Correct improper biomechanics and technique
  - 🕮 Equipment modifications (increase handle diameter and decrease string tension for tennis rackets)

- *Surgical:*
  - ECR-B debridement
- 🕮 *Posttreatment* return to play, a tennis player should:
  - Decrease string tension
  - Increase grip size

OLECRANON BURSITIS (FIGURE 4–42)

General

- Also known as draftsman's elbow, student's elbow, or miner's elbow

MECHANISM

- Repetitive trauma, inflammatory disorder (gout, pseudogout, rheumatoid arthritis [RA])

PATHOLOGY

- Inflammation of the bursa located between the olecranon and skin

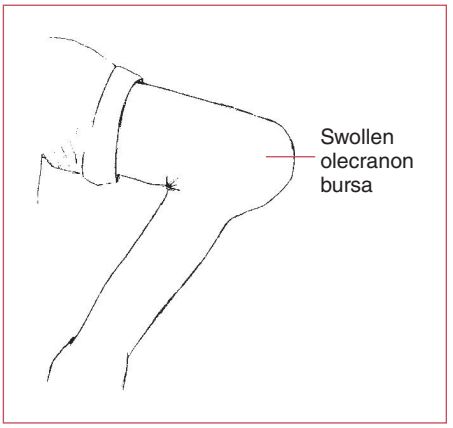

FIGURE 4–42 Olecranon bursitis.

Clinical Features

- Swelling and pain in the posterior aspect of the elbow and decreased elbow ROM
- A warm, erythematous elbow may indicate infection.

Imaging

- None needed

Treatment

- Fluid aspiration and culture if indicated
- *Conservative:* Rest, NSAIDs, elbow padding

DISLOCATION OF THE ELBOW

General

- The most common type of dislocation in children and the second most common type in adults (second only to shoulder dislocation)
- Young adults 25 to 30 years old are most affected and sports activities account for almost 50% of these injuries.

MECHANISM OF INJURY

- Fall on an outstretched hand

Clinical Features

- Dislocation can be anterior or posterior, with *posterior* being the most common, occurring 98% of the time **(Figure 4–43)**.
- Associated injuries include fracture of the radial head, as well as injury to the brachial artery and median nerve.

SYMPTOMS

- Inability to bend the elbow following a fall on the outstretched hand
- Pain in the shoulder and wrist
- The most important part of the exam is the neurovascular evaluation of the radial artery, and the median, ulnar, and radial nerves.

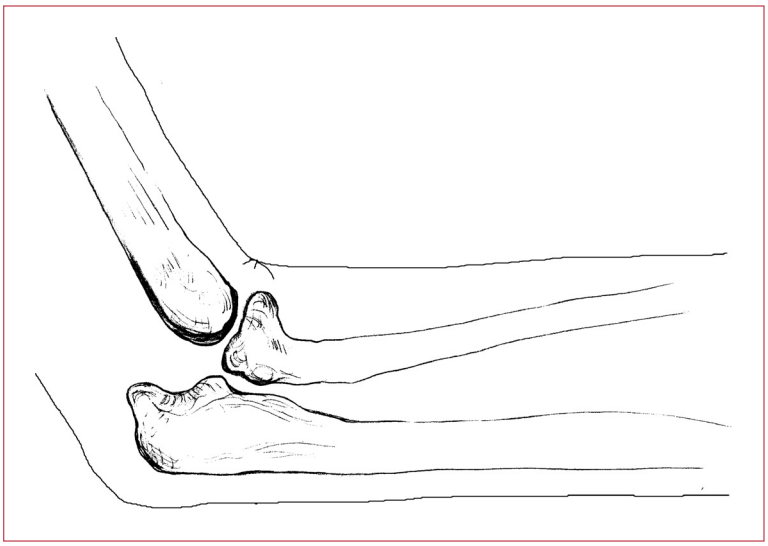

FIGURE 4–43 Posterior dislocation of the elbow.

Imaging

- Plain AP and lateral radiographs
- CT and MRI scans are seldom necessary.

Treatment

- Reduce dislocation as soon as possible after injury.
- Splint for 10 days.
- Initiate ROM exercises and NSAIDs.

Adverse Outcomes

- Loss of ROM of elbow, especially extension
- Ectopic bone formation
- Neurovascular injury
- Arthritis of the elbow

DISTAL BICEPS TENDONITIS

General

- Overloading of the biceps tendon, commonly due to repetitive elbow flexion and supination or resisted elbow extension

PATHOLOGY

- Microtearing of the distal biceps tendon

COMPLICATION

- Biceps tendon avulsion

Clinical Features

- Insidious onset of pain in the antecubital fossa usually after an eccentric overload
- Audible snap with an obvious deformity ("Popeye sign"), swelling, and ecchymosis if an avulsion is suspected

Imaging
- None needed

Treatment
- *Conservative:*
  - Relative rest, ice, NSAIDs
  - Physical therapy modalities
  - Correct improper technique
- *Surgical:*
  - Reattachment if there is tendon rupture/avulsion

TRICEPS TENDONITIS/AVULSION

General
- Tendonitis: Overuse syndrome secondary to repetitive elbow extension
- Avulsion: Decelerating counterforce during active elbow extension

Clinical Features
- Posterior elbow pain with tenderness at the insertion of the triceps tendon
- Pain with resistive elbow extension
- Sudden loss of extension with a palpable defect in the triceps tendon (avulsion)

Imaging
- Plain films to rule out other causes if indicated

Treatment
- Conservative
- *Surgical:* Reattachment

VALGUS EXTENSION OVERLOAD SYNDROME OF THE ELBOW

General
- Spectrum of overuse elbow injuries in baseball players caused by repetitive valgus forces during the throwing motion, especially in cocking and acceleration phases of throwing
- Valgus forces cause tensile stress in the medial elbow and lateral shear stress in the posterior aspect of the elbow (posteromedial olecranon).

PATHOLOGY
- Olecranon osteophytosis and loose body formation occur secondary to repetitive abutment of the olecranon against the olecranon fossa.

Clinical Features
- Posterior elbow pain with lack of full elbow extension
- Catching or locking during elbow extension
- 🕮 **Provocative test: Valgus extension overload (VEO) test**
  - Flex elbow to 30 degrees and repeatedly extend the elbow fully while applying a valgus stress.
  - Pain may be elicited, particularly at the last 5 to 10 degrees of extension.
  - Valgus stress test should also be performed at >90 degrees to rule out UCL injury.

Imaging
- AP/lateral x-rays may show a loose body or osteophyte formation at the olecranon.

Treatment

- Surgical removal of the loose body/osteophyte
- Postoperative physical therapy focuses on stretching and strengthening eccentric elbow flexors to better control rapid elbow extension, as well as evaluation of pitching biomechanics.

MEDIAL (ULNAR) COLLATERAL LIGAMENT SPRAIN

General

- A repetitive valgus stress occurring across the elbow, most prominently during the acceleration phase of throwing

PATHOLOGY

- Inflammation of the anterior band of the UCL, which is the segment that provides the majority of valgus stability

Clinical Features

- Significant medial elbow pain occurring after the throwing motion
- A pop or click may be heard precipitating the pain.
- Medial pain or instability on valgus stress with the elbow, flexed 20 to 30 degrees if the UCL is torn

PROVOCATIVE TEST: VALGUS STRESS TEST

- Tenderness over the medial aspect of the elbow, which may be increased with a valgus stress
- Should perform VEO test to differentiate between UCL injury and VEO syndrome

Imaging

- Plain films may reveal calcification and spurring along the UCL.
- Valgus stress radiographs demonstrate a 2-mm joint space suggestive of UCL injury.
- On US, applying a valgus force during examination may show increased joint space.

Treatment

- *Conservative:*
  - Rest, ice, NSAIDs
  - Rehabilitation program for strengthening and stretching
  - Establishing return-to-play criteria
- *Surgical* reconstruction if needed

LATERAL (RADIAL) COLLATERAL LIGAMENT SPRAIN

General

- Usually associated with a traumatic elbow dislocation

Clinical Features

- *Recurrent* locking or clicking of the elbow with extension and supination
- *Lateral* elbow pain or instability on varus stress with the elbow flexed 20 to 30 degrees if the LCL is torn

PROVOCATIVE TESTS

- **Varus stress test:**
  - Tenderness over the lateral aspect of the elbow, which may be increased with a varus stress
- **Lateral pivot-shift test:**
  - Assesses the LCL for posterolateral instability.

Imaging

- **Varus** stress radiographs demonstrating a 2-mm joint space are suggestive of LCL injury.

Treatment

- *Conservative:*
  - Rest, ice, NSAIDs
  - Rehabilitation program for strengthening and stretching
  - Establishing return-to-play criteria
- *Surgical* reconstruction if needed

PRONATOR SYNDROME (ALSO SEE CHAPTER 5, ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY)

General

- 🕮 Median nerve compression at the elbow by the following structures:
  - Ligament of Struthers or supracondylar spur
  - Lacertus fibrosus
  - PT muscle
  - Between the two heads of the FDS

Clinical Features

- Dull aching pain in the proximal forearm just distal to the elbow
- Numbness in the median nerve distribution of the hand
- Symptoms exacerbated by pronation

Imaging

- Plain films: Rule out bone spur.
- Electromyography/nerve conduction studies (EMG/NCS) to assess for median neuropathy at the elbow

Treatment

- *Conservative:*
  - Modification of activities
  - Avoid aggravating factors
  - Stretching and strengthening program
- *Surgical:* Release of the median nerve at the location of the compression

CUBITAL TUNNEL SYNDROME (ALSO SEE CHAPTER 5, ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY)

General

- A number of factors can compromise the integrity of the ulnar nerve in the region of the elbow:
  - Arcade of Struthers
  - Hypermobility of the ulnar nerve
  - Excessive valgus force at the elbow
  - Impingement from osteophytes or loose bodies

PATHOLOGY

- Hyperirritability or injury of the ulnar nerve

Clinical Features

- Medial forearm aching pain with paresthesias radiating distally to the fourth and fifth digits
- Weakness in the ulnar-innervated hand intrinsic musculature: Weak grip strength, muscle atrophy

- Positive Tinel's sign at the elbow
- Positive Froment's sign

Imaging

- X-ray to evaluate for osteophytes or loose bodies
- Consider MRI for soft tissue abnormalities if indicated
- EMG/NCS above and below the elbow

Treatment

- *Conservative:* Relative rest, NSAIDs, elbow protection (splinting), and technique modification
- *Surgical:* Ulnar nerve transposition

OSTEOCHONDROSIS OF THE ELBOW (PANNER'S DISEASE)

General

- Epiphysial aseptic necrosis of the capitellum
- Should not be confused with osteochondritis dissecans of the capitellum of the elbow (localized fragmentation of the bone and cartilage of the capitellum)

MECHANISM

- Believed to be caused by interference in blood supply to epiphysis, leading to resorption of the ossification center initially, followed by repair/replacement

Clinical Features

- Symptoms relieved by rest and aggravated by activity
- Tenderness and swelling on the lateral aspect of the elbow
- Usually seen in dominant elbow of young boys
- Limited extension seen on ROM

Imaging

- Plain films: Sclerosis, patchy areas of lucency with fragmentation

Treatment

- *Conservative:* Immobilization, then gradual ROM

FRACTURE OF THE HUMERAL SHAFT

General

- Fairly common—constituting up to 5% of all fractures

MECHANISM

- Direct trauma (e.g., MVA)
- Fall on outstretched arm

Clinical Features

- Severe arm pain and swelling and deformity are characteristic of a displaced fracture of the humerus.
- 🕮 If the radial nerve has been injured, patients may exhibit weakness of radial nerve innervated muscles with sparing of the triceps **(Figure 4–44)**.

FIGURE 4–44 Radial nerve entrapment at the humeral shaft fracture site.

Imaging

- AP and lateral x-rays to confirm diagnosis

Treatment

- Humeral shaft fractures can be treated conservatively (splint for 2 weeks).
- 🕮 Special problem associated with humeral shaft fracture is radial nerve injury.
- 95% of patients will regain their nerve function within 6 months. During this period of observation, the patient should wear a splint and work with a therapist.
- EMGs are indicated if radial nerve function does not return.

FRACTURE OF THE DISTAL HUMERUS

General

Classification can be complex. The most useful way to consider them is displaced or nondisplaced. A displaced fracture involves one or both condyles, and the joint surface may or may not be involved (**Figure 4–45**):

- Complications:
  - Neurovascular injury
  - Nonunion
  - Malunion
  - Elbow contracture
  - Poor ROM

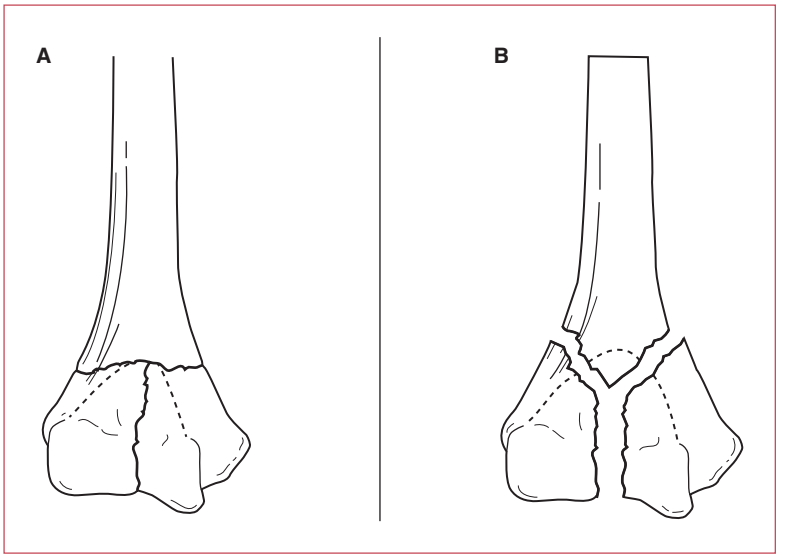

FIGURE 4–45 (A) Distal humerus: Nondisplaced condylar fracture. (B) Distal humerus: Displaced intercondylar fracture.

Clinical Features

The patient will demonstrate swelling, ecchymosis, and pain at the elbow:

- Inability to flex the elbow
- Inspect for an obvious deformity.
- Neurovascular compromise: Radial, median, and ulnar nerves all may be affected.

Imaging

- AP/lateral x-rays of the elbow

Treatment

- Orthopedic referral:
  - Nondisplaced fractures can be treated by splinting and early motion.
  - Displaced fractures—except severely comminuted fractures—require open reduction with fixation.

RADIAL HEAD FRACTURE

General

- Dislocations of the elbow are commonly associated with radial head fractures.

CLASSIFICATION (FIGURE 4–46)

Type I	Nondisplaced or minimally displaced <2 mm or marginal lip fracture
Type II	Fracture displaced >2 mm or fracture involving more than marginal lip
Type III	Comminuted fracture

Sources: From Iannuzzi NP, Leopold SS. In brief: the Mason classification of radial head fractures. *Clin Orthop Relat Res*. 2012;470(6):1799–802. doi:10.1007/s11999-012-2319-2. Epub 2012 Mar 17. PMID: 22426693; PMCID: PMC3348319; Al-Tawil K, Arya A. Radial head fractures. *J Clin Orthop Trauma*. 2021;20:101497. doi:10.1016/j.jcot.2021.101497. PMID: 34307018; PMCID: PMC8283329.

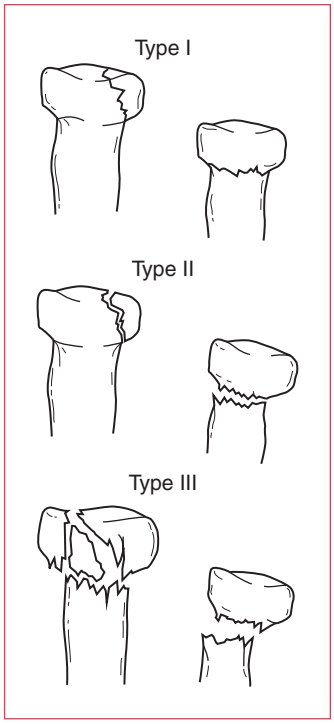

FIGURE 4–46 Radial head fracture classification.

Clinical Features

- Fall on an outstretched arm, causing pain, swelling, and ecchymosis around the elbow
- Pain and decreased ROM observed in elbow flexion and extension, pronation, and supination

IMAGING

- Plain films of the elbow

Treatment

- Orthopedic referral:
  - **Type I** (nondisplaced):
    - *Conservative:* Short period of immobilization (3–5 days) followed by early ROM
  - **Type II** (minimal displacement):
    - Surgical fixation for fracture >2 mm displacement or 30% radial head involvement
  - **Type III** (comminuted fracture):
    - Surgical fixation

OLECRANON FRACTURE

General

- Direct blow to the elbow, such as a fall onto the elbow with the elbow flexed
- Fall on an outstretched arm in association with a dislocation
- 🕮 Most common stress fracture type among baseball players
- Pain localized to the posterior lateral border of the ulna at the olecranon articular surface
- Pain with acceleration phase of throwing

CLASSIFICATION
- Nondisplaced
- Displaced

Clinical Features
- Swelling and ecchymosis with an obvious deformity
- Pain on gentle ROM
- Numbness and paresthesias with radiation distally to the fourth and fifth digits with ulnar nerve involvement

Imaging
- Plain films: AP, lateral, and oblique

Treatment
- Nondisplaced: Conservative (immobilization followed by physical therapy)
- Displaced: Surgical fixation

🕮 NIGHTSTICK FRACTURE (FIGURE 4–47)

General
- Isolated fracture of the ulnar shaft
- Name is derived from the idea of a suspect holding up their arm while being hit by a police nightstick
- Typically from a direct blow; can also result from indirect trauma such as a mechanical fall

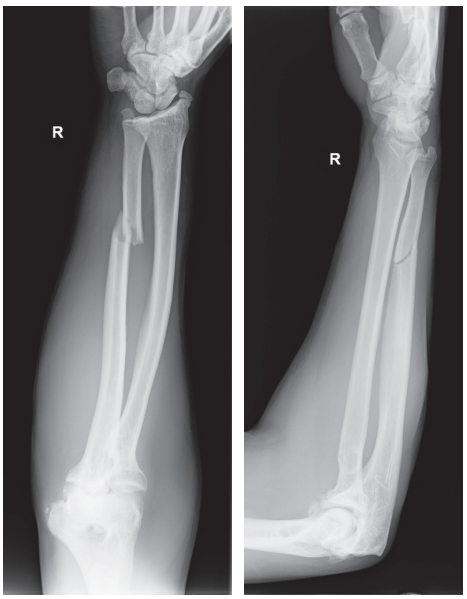

FIGURE 4–47 Nightstick fracture.

CLASSIFICATION
- Type A—Simple fracture
- Type B—Wedge fracture
- Type C—Comminuted (Multifragment) fracture

Clinical Features
- Pain and swelling
- Possible loss of forearm and hand function
- Typically closed fractures

Imaging
- Plain films of the elbow

Treatment
- <50% displaced: Nonoperative management with cast, bracing, or soft compression dressings
- >50% displaced or proximal one-third isolated ulna fracture: Surgical fixation

🕮 MONTEGGIA FRACTURE-DISLOCATION (FIGURE 4–48)

General
- Defined as a proximal one-third ulnar fracture associated with radial head dislocation
- Rare in adults; more common in children, with different treatment protocols in children

CLASSIFICATION

- Bado classification focuses primarily on the radial head.
  - Type I: Anterior dislocation of the radial head (most common type)
  - Type II: Posterior dislocation
  - Type III: Lateral dislocation
  - Type IV: Dislocation in any direction associated with both radial and ulnar shaft fractures

FIGURE 4–48 Monteggia fracture.

Clinical Features

- Pain and swelling at elbow joint
- May have deformity and obvious dislocation of the radiocapitellar joint
- Can be associated with posterior interosseous nerve (PIN) neuropathy

Imaging

- Elbow and forearm radiographs

Treatment

- Most adults with Monteggia fractures require surgical fixation.
- Children with stable elbow joint and anatomic alignment are typically treated with closed reduction and casting.

GALEAZZI FRACTURE (FIGURE 4–49)

General

- Defined as distal one-third radial shaft fracture that is also associated with distal radioulnar joint (DRUJ) instability
- Result from direct wrist trauma, typically on the dorsolateral aspect of the wrist
- Fall onto outstretched hand with forearm in pronation

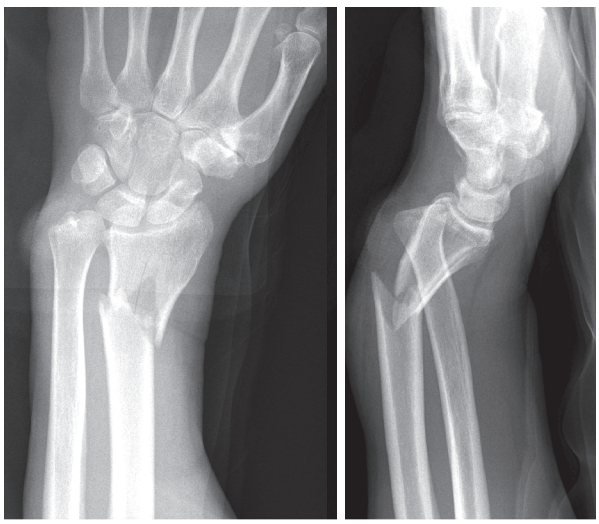

FIGURE 4–49 Galeazzi fracture.

Clinical Features

- Pain, swelling, deformity of the forearm and wrist
- DRUJ stress causes wrist or midline forearm pain.

Imaging

- AP and lateral plain films of the elbow, forearm, and wrist

Treatment

- ORIF of radius with reduction and stabilization of the DRUJ in all cases

UPPER EXTREMITIES: THE WRIST REGION

FUNCTIONAL ANATOMY

Ranges of Motion at the Wrist (Figure 4–50)

Wrist flexion	80 degrees
Wrist extension	70 degrees
Ulnar deviation of the wrist	35–40 degrees
Radial deviation of the wrist	20–25 degrees

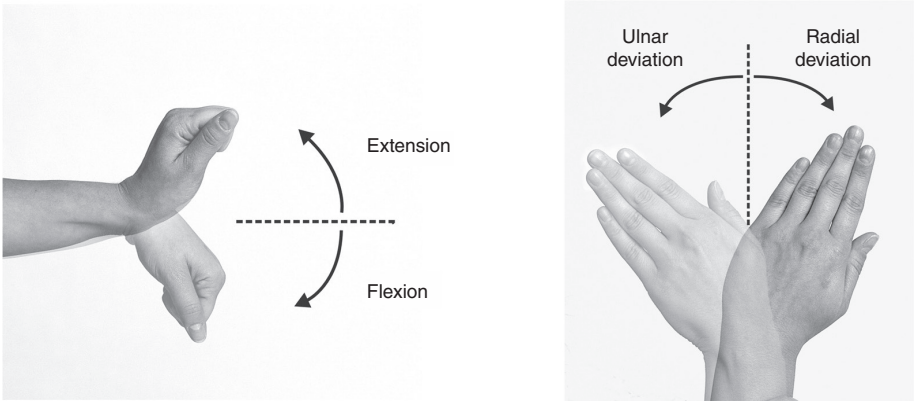

FIGURE 4–50 Wrist range of motion terminology.

Carpal Bones (Figure 4–51)

- **Proximal row: "Some Lovers Try Positions" (radial → ulnar direction):**
  - Scaphoid
  - Lunate
  - Triquetrum
  - Pisiform
- **Distal row: "That They Can't Handle" (radial → ulnar direction):**
  - Trapezium
  - Trapezoid
  - Capitate
  - Hamate

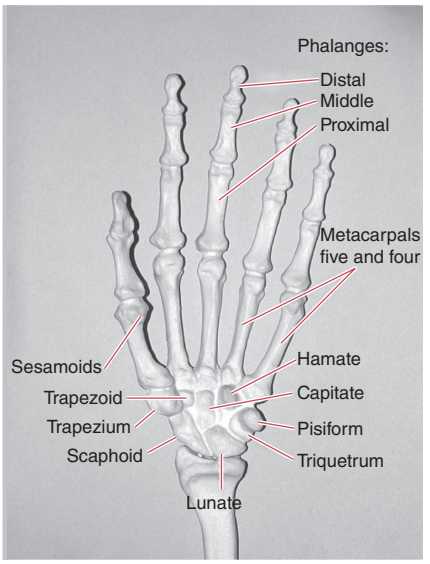

FIGURE 4–51 Palmar view—bones of the wrist and hand.

Wrist Flexors (Figure 4–52)

WRIST FLEXORS	SPINAL INNERVATION	PERIPHERAL INNERVATION
FCR	C6, C7	Median nerve from median + lateral cords
FCU	C8, T1	Ulnar nerve from medial cord
Palmaris longus	C7, C8	Median nerve from medial + lateral cords
FDS	C7, C8, T1	Median nerve from medial + lateral cords
FDP	C7, C8, T1	Median nerve from medial + lateral cords to second and third digits; Ulnar nerve from medial cord: fourth and fifth digits
FPL	C8, T1	Median nerve from medial + lateral cords

FCR, flexor carpi radialis; FCU, flexor carpi ulnaris; FDP, flexor digitorum profundus; FDS, flexor digitorum superficialis; FPL, flexor pollicis longus.

Wrist Extensors (Figure 4–53)

WRIST EXTENSORS	SPINAL INNERVATION	PERIPHERAL INNERVATION
ECR-L	C6, C7	Radial nerve from posterior cord
ECR-B	C6, C7	Radial nerve from posterior cord
ECU	C7, C8	Radial nerve from posterior cord
EDC	C7, C8	Radial nerve from posterior cord
EPL	C6, C7, C8	Radial nerve from posterior cord
Extensor indicis	C6, C7, C8	Radial nerve from posterior cord
EDM	C8, T1	Ulnar nerve from medial cord

ECR-B, extensor carpi radialis brevis; ECR-L, extensor carpi radialis longus; ECU, extensor carpi ulnaris; EDC, extensor digitorum communis; EDM, extensor digitorum minimi; EPL, extensor pollicis longus.

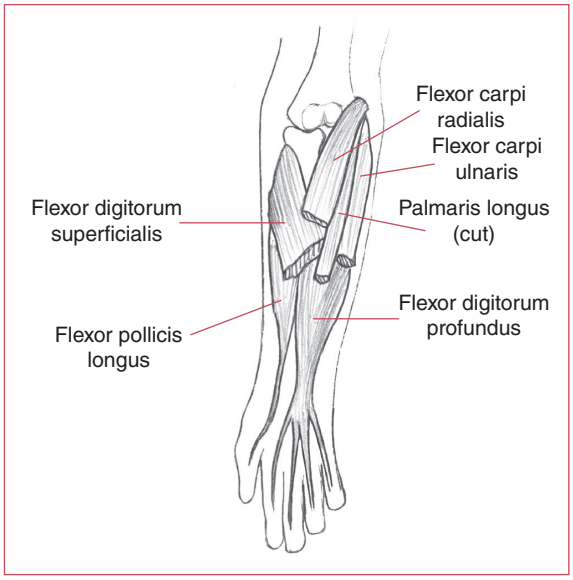

FIGURE 4–52 Wrist flexors.

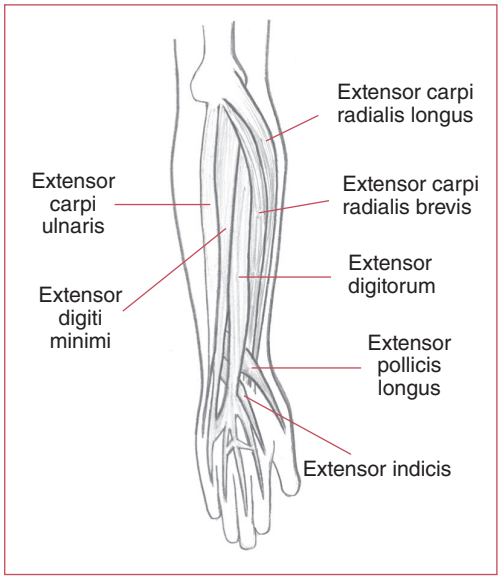

FIGURE 4–53 Wrist extensors.

Ulnar Deviation of the Wrist (Adduction)

ULNAR DEVIATION	SPINAL INNERVATION	PERIPHERAL INNERVATION
ECU	C7, C8	Radial nerve from posterior cord
FCU	C8, T1	Ulnar nerve from medial cord

ECU, extensor carpi ulnaris; FCU, flexor carpi ulnaris.

Radial Deviation of the Wrist (Abduction)

RADIAL DEVIATION	SPINAL INNERVATION	PERIPHERAL INNERVATION
FCR	C6, C7	Median nerve from medial + lateral cords
ECR-L	C6, C7	Radial nerve from posterior cord

ECR-L, extensor carpi radialis longus; FCR, flexor carpi radialis.

Extensor Compartments of the Wrist (Figure 4–54)

- **First compartment:**
  - Abductor pollicis longus (APL—"All peanut lovers")
  - Extensor pollicis brevis (EPB—"Eat peanut butter")
- **Second compartment:**
  - ECR-L
  - ECR-B
- **Third compartment:**
  - Extensor pollicis longus (EPL)
- **Fourth compartment:**
  - Extensor digitorum communis (EDC)
  - Extensor indices proprius (EIP)
- **Fifth compartment:**
  - Extensor digitorum minimi (EDM)
- **Sixth compartment:**
  - ECU

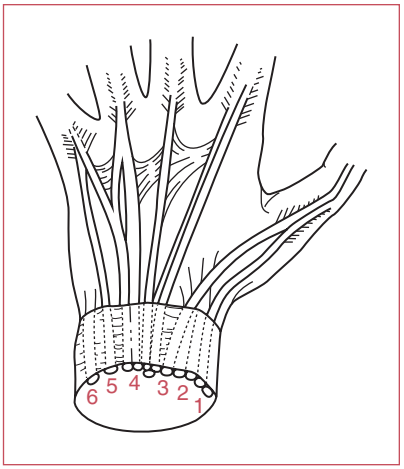

FIGURE 4–54 Extensor tendons with the six tendon sheath compartments (dorsum of the wrist).

WRIST DISORDERS

ARTHRITIS

General

TYPES

- **OA**: Noninflammatory disorder with deterioration of the articular cartilage and formation of new bone at the joint margins
- **RA**: Autoimmune attack on the synovial tissue, destroying the articular cartilage, leading to bone destruction. See the Rheumatoid Arthritis section in Chapter 3, Rheumatology, for a detailed discussion.

Clinical Features

- 🕮 **OA:**
  - Heberden's and Bouchard's nodules involve the distal interphalangeal (DIP) and proximal interphalangeal (PIP) joints, respectively.

  - Tenderness along the area of involvement and crepitus with wrist ROM:
    - Common in the first carpometacarpal (CMC) joint of the thumb
    - For testing CMC joint involvement, axial compression of the metacarpal on the trapezium gives a painful grinding sensation. The grind test identifies mild to severe disease. There may be localized tenderness over the ulnar aspect of the thumb.
  - Cyst formation occurs in the joint space.
- **RA:**
  - Synovitis in the hands/wrists primarily affecting the metacarpophalangeal (MCP) and PIP joints
  - Ulnar deviation of the MCPs
  - Radial deviation of the wrist
  - Dorsal subluxation of the ulna
  - Erosion of the ulnar styloid at the end stage
  - *Swan neck deformity:*
    - Caused by shortening and contracture of the intrinsic muscles of the hand
    - Flexion at the MCP joint
    - Hyperextension at the PIP joint
    - Flexion at the DIP joint
  - *Boutonnière deformity:*
    - Caused by tearing of the extensor hood
    - Hyperextension at the MCP joint
    - Flexion at the PIP joint
    - Hyperextension of the DIP joint

Imaging

- Plain films of the wrist and digits

Treatment

- *Conservative*. See the Rheumatoid Arthritis section in Chapter 3, Rheumatology, for a detailed discussion.

DE QUERVAIN'S TENOSYNOVITIS

General

- Repetitive or direct trauma to the sheath of the EPB and APL tendons, causing a tenosynovitis and inflammation
- Involvement of the tendons in the first compartment of the wrist

Clinical Features

- Pain and tenderness on the radial side of the wrist associated with movement
- Edema and crepitus may also be present.

PROVOCATIVE TEST

- **Finkelstein's test (Figure 4–55):**
  - Flex the thumb into the palm of the hand with the fingers, making a fist over the thumb. Then passively ulnar deviate the wrist.
  - Pain elicited with a positive test
  - May also be positive in patients with RA

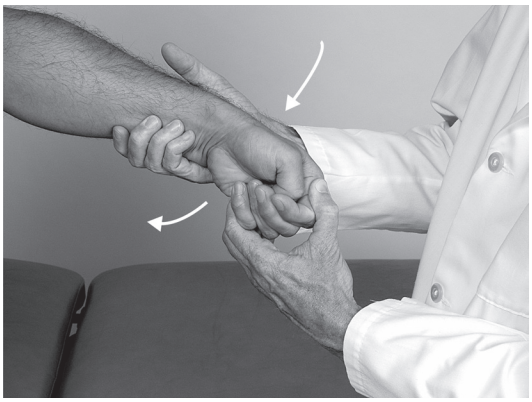

FIGURE 4–55 Finkelstein's test.
Source: Courtesy of JFK Johnson Rehabilitation Institute, 2000.

Imaging

- None needed

Treatment

- *Conservative:*
  - Thumb spica splint to immobilize the thumb
  - NSAIDs
  - Corticosteroid injection
- *Surgical* release of the tight sheath eliminates the friction that worsens the inflammation, thus restoring the tendon's smooth gliding capability.
- 🕮 For persistent or recurrent symptoms >6 months, surgical decompression of the first extensor compartment is indicated (Fam, 2007).

GANGLION CYST (FIGURE 4–56)

General

- Synovial fluid-filled cystic structure that arises from the synovial sheath of the joint space

Clinical Features

- Small smooth mass on the dorsal or volar aspect of the wrist that occurs on the dorsal aspect in 60% of cases
- Pain may occur with ranging the wrist or slight pressure.

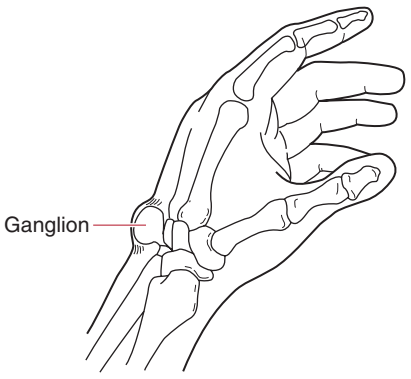

FIGURE 4–56 Wrist ganglion.

Imaging

- Plain films of the wrist if indicated

Treatment

- Immobilization
- Aspiration of the cyst (90% recurrence)
- Surgical removal if needed (10% recurrence)

OSTEONECROSIS OR AVASCULAR NECROSIS OF THE LUNATE (FIGURE 4–57)

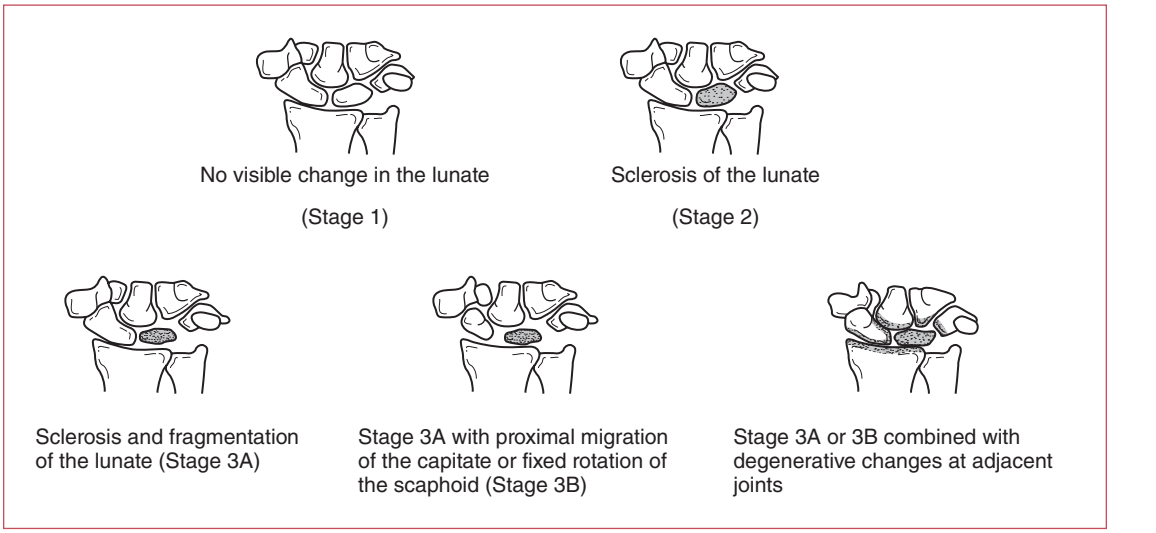

FIGURE 4–57 Kienböck's disease classification.

General

- 🕮 Also known as Kienböck's disease

MECHANISM OF INJURY

- Idiopathic loss of blood supply to the lunate, which causes AVN of the bone
- Thought to be caused by vascular impairment and/or repeated trauma (repeated stress or fracture)
- Bone collapse results in degenerative changes at the wrist.

RISK FACTORS

- Poor vascular supply to the area
- Short ulnar variance:
  - Patients with a short ulna are thought to have an increased incidence of osteonecrosis of the lunate as compared with normal individuals because of the increased shear forces that are placed on the lunate.

Clinical Features

- Ulnar-sided pain, stiffness, and swelling over the dorsal aspect of the wrist directly over the lunate
- Reduced grip strength

Imaging

- Plain films: May see a compression fracture, flattening, or sclerosis of the lunate
- Bone scan: Increased uptake of the lunate
- MRI: Increased signal intensity on T2, decreased on T1, of the lunate

Treatment

- Orthopedic referral

SCAPHOID FRACTURE

General

- One of the most common fractures of the wrist, comprising 70% of all carpal bone fractures

MECHANISM OF INJURY

- A fall or blow on a hyperextended (dorsiflexed) wrist
- Osteonecrosis of the bone may develop secondary to its blood supply.
- The majority of the blood supply is to the distal one-third of the bone. Therefore, the middle and proximal portions of the bone have a large nonunion rate (one-third developing osteonecrosis).

Classification: Anatomic Location (Figure 4–58)

- Waist (65%)
- Tubercle (2%)
- Distal pole (10%)
- Proximal pole (15%)

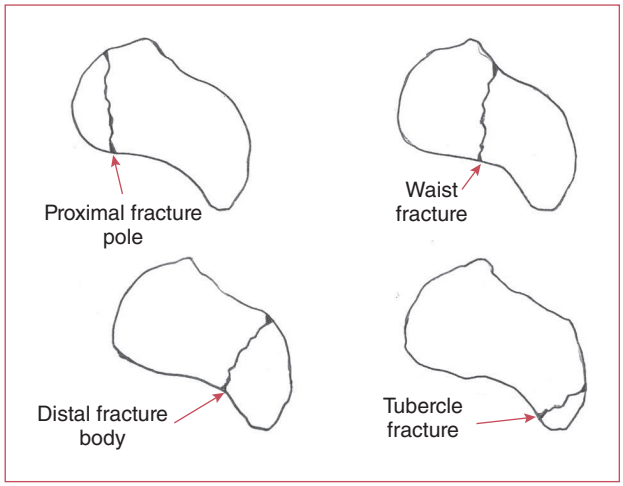

FIGURE 4–58 Anatomic location of scaphoid fractures.

Complications

- Osteonecrosis, which may lead to carpal bone collapse (scapholunate) if not properly diagnosed and treated

🕮 Clinical Features

- Swelling and tenderness in the areas of the thumb and wrist (anatomic snuff box)
- Pain with ROM, especially in extension and radial deviation
- Tenderness to palpation over the tuberosity of the scaphoid
- Anatomic snuff box: Borders (**Figure 4–59**):
  - Base: Scaphoid bone
  - Lateral: APL and EPB
  - Medial: EPL

FIGURE 4–59 Anatomic snuffbox.

Imaging

- Plain films: Posterior-anterior (PA) and oblique view of the wrist in ulnar deviation with comparisons to the opposite side if needed. Repeat in 2 weeks if no fracture is seen initially.
- Repeat films at 4 to 6 weeks if still symptomatic.
- CT scan can be done if there is a question of fracture.
- Bone scan can be positive as early as 24 hours after injury.

Treatment

- A fracture may or may not be visualized on initial x-ray imaging. Therefore, a patient with tenderness in the area of the anatomic snuff box has a fracture until proven otherwise and should be treated accordingly.
- Immobilize the wrist in a thumb spica cast for 10 to 14 days and repeat the radiographs.
- Scaphoid fractures are classified in multiple ways, most commonly based on location or stability of the fracture.
- The location of the fracture (10% proximal pole, 70% waist, 20% distal pole), stability of fracture, and the timing of injury will dictate treatment.
- Conservative management:
  - Immobilization of the wrist in a long thumb spica cast for 6 weeks with the wrist in a neutral position
  - At 6 weeks, change to a short thumb spica cast if the plain films show proper healing.
  - If poor healing occurs at this time, surgical stabilization may be indicated.
- Potential surgical intervention:
  - Fractures of the proximal pole, displaced fractures >1 mm, and fractures with delayed presentation (<2–3 weeks to diagnose) should be referred to orthopedics due to the high risk of AVN.
- On average
  - Distal pole fractures take 8 weeks to heal.
  - Waist (middle portion) fractures (most common) take 3 months to heal.
  - Proximal pole fractures take 4 months or longer to heal.

🕮 HAMATE FRACTURES

General

- Body fractures often happen from direct trauma; hamate fractures can occur involving axial loading through the fourth or fifth metacarpal.
- 🕮 Fractures of the hook of the hamate can occur as a result of direct trauma on the palmar surface of the wrist/hand or due to avulsion from shear forces from adjacent/attached tendons during forceful twisting motion of the wrist.

- Sports activities commonly related to this injury mechanism include racquet-related sports, baseball, and golf, with injurious forces occurring at end swing.
- Located slightly distal and radial to the pisiform and forms radial border of the tunnel of Guyon, through which the ulnar nerve traverses
- Vascular supply at radial base and ulnar tip

Clinical Features

- Ulnar and palmar wrist pain with racquet, bat, or club swing
- Pain over the hook of the hamate, and/or pain over the dorsal ulnar hamate
- Injury to the ulnar nerve may lead to accompanying paresthesias or weakness in ulnar distributions.
- Pain worse with axial loading of the fourth and fifth metacarpals
- Provocative maneuver of resisted finger flexion with the wrist held in ulnar deviation may be painful.

Imaging

- X-rays with PA, lateral, carpal tunnel, and 45-degree supinated oblique views
- CT may be needed for fractures at the base of the hook.

Treatment

- Nondisplaced body fractures: 4 to 6 weeks short arm cast
- Displaced body fractures require surgical referral.
- Acute hook fractures: 50% heal after prolonged casting (6 weeks to 4 months).
- Most acute hook fractures should have adequate vascular supply to heal if immobilized within the first week.
- Cast should maintain slight wrist flexion and slight MCP flexion to reduce shear forces from fourth and fifth digit flexor tendons.
- Injuries older than 2 weeks may require excision.
- Persistent numbness of ulnar digits requires electrodiagnostic evaluation.

TRAPEZIUM FRACTURES

General

- Isolated fractures are rare.
- May occur when the base of the thumb metacarpal is forced by axial load into the trapezium
- Can also occur as an avulsion fracture

TYPES

- Body
- Trapeziometacarpal
- Trapezial ridge

Clinical Features

- Body and trapeziometacarpal fractures present as tenderness of the dorsal wrist proximal to the thumb CMC joint.
- Trapezial ridge fractures present as a tender point distal to the scaphoid tuberosity.

Imaging

- X-rays with PA, lateral, oblique, carpal tunnel, and Bett's views (thumb extended and abducted, wrist slightly pronated, beam centered on the scaphotrapeziotrapezium area)
- Trapeziometacarpal

Treatment

- Nondisplaced body, proximal trapezial ridge fractures, and trapeziometacarpal fractures require a short arm thumb spica cast for 4 weeks.
- Displaced body and distal trapezial ridge fractures require surgical referral.

FRACTURES OF THE DISTAL RADIUS

General

Fractures are named according to the displacement and angulation of the distal bone:

- **Colles' fracture (Figure 4–60A):**
  - Most common type of fracture
  - Fracture of the distal radius with dorsal displacement and angulation
  - Associated with triangular fibrocartilage complex tears and scapholunate dissociation
- **Smith's fracture (Figure 4–60B):**
  - Fracture of the distal radius with volar displacement and angulation
  - Reverse of Colles' fracture

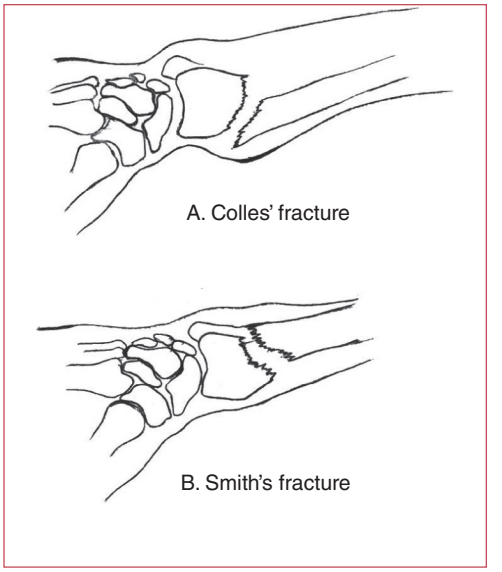

FIGURE 4–60 (A) Colles' fracture (note radial dorsal angulation). (B) Smith's fracture.

Clinical Features

- Acute pain, swelling at the wrist, usually after a fall on an outstretched arm

Imaging

- AP and lateral plain films of the wrist and hand

Treatment

- Orthopedic referral for closed reduction depending on the location, degree of displacement, and reproducibility

UPPER EXTREMITIES: THE HAND REGION

FUNCTIONAL ANATOMY

Bones of the Hand (Figure 4–61)

- Eight carpal bones (see also Upper Extremities: The Wrist Region section)
- Five metacarpals
- Fourteen phalanges:
  - Five proximal
  - Four middle (not located in the thumb)
  - Five distal

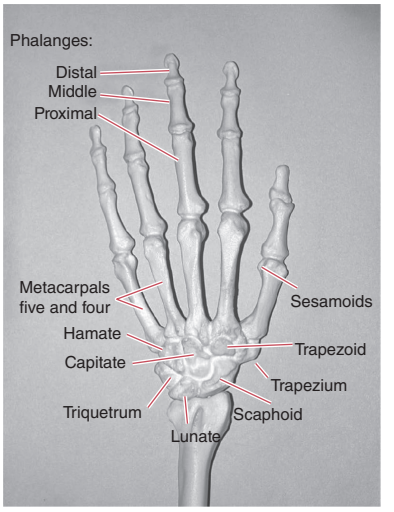

FIGURE 4–61 Dorsal view—bones of the wrist and hand.

Tendon Function: "Pulley System"

- Extensor system:
  - EDC inserts in the terminal portions of digits 2 to 5
- Flexor system:
  - Two-tendon pulley system made up of the FDP and FDS (Figure 4–62)

Ranges of Motion of the Digits (Figure 4–63)

- **Flexion:**
  - MCP: 90 degrees
  - PIP: 90 degrees

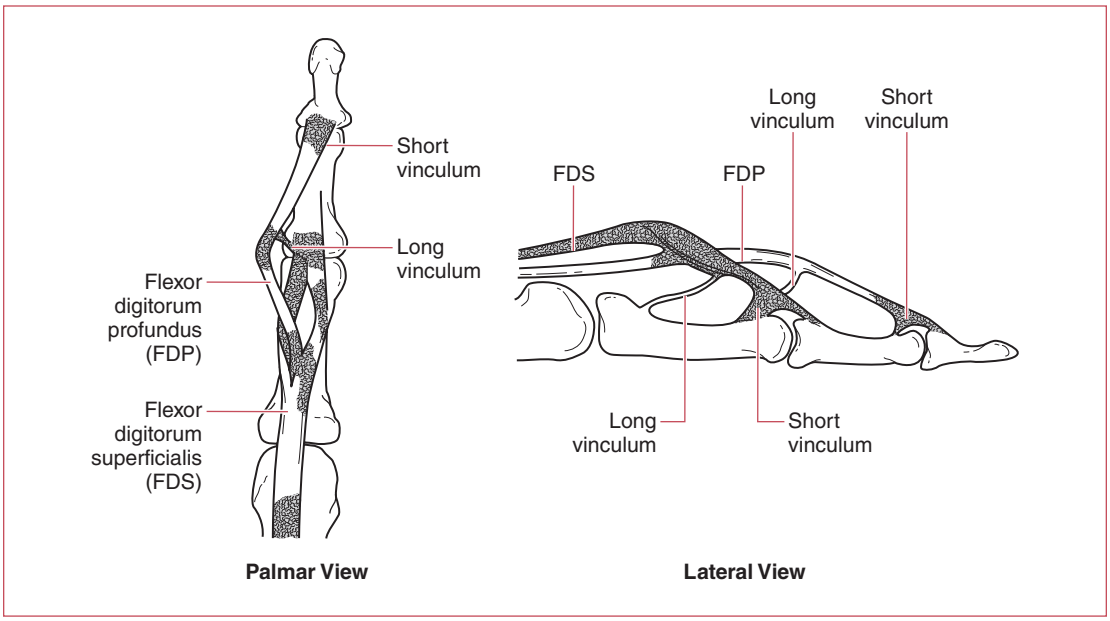

FIGURE 4–62 Flexor tendon system.

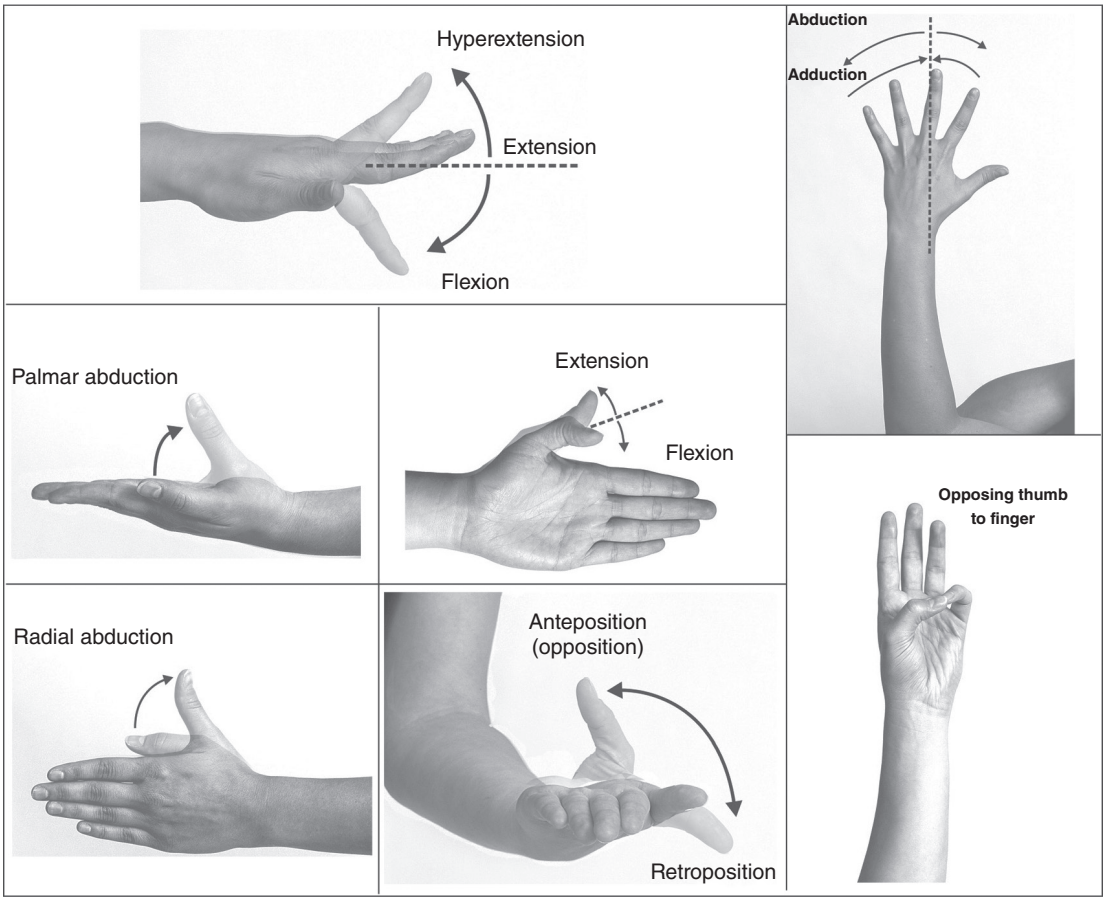

FIGURE 4–63 Finger and thumb range of motion terminology.

  - DIP: 90 degrees
  - Thumb: MCP 50 degrees, interphalangeal (IP) 90 degrees
- **Extension:**
  - MCP: 30 degrees
  - PIP: 0 degrees
    - DIP: 0 to 10 degrees
    - Thumb: MCP 0 degrees, IP 20 degrees
- **Abduction:**
    - Finger: 20 degrees – Thumb: 70 degrees
- **Adduction:**
    - Finger: 0 degrees (return from abduction)
    - Thumb: 0 degrees (return from abduction)
- **Opposition:**
    - Thumb: Approximation of the palmar aspect of the thumb and fifth digit
- **Apposition:**
    - Thumb: Approximation between the thumb and other digit not using the palmar aspect

Digit Motion (Figures 4–64 to 4–66)

- **Finger flexors:**
  - FDP:
    - Digits two and three (anterior interosseous nerve [AIN; median nerve]: C8, T1)
    - Digits four and five (ulnar nerve: C8, T1)
  - FDS (median nerve: C7, C8, T1)
  - Lumbricals:
    - First and second lumbricals (median nerve: C8, T1)
    - Third and fourth lumbricals (ulnar nerve: C8, T1)
  - Dorsal and palmar interossei (ulnar nerve: C8, T1)
  - Flexor digiti minimi (ulnar nerve: C8, T1)
- **Finger extensors:**
  - EDC (PIN [radial nerve]: C7, C8)
  - Extensor indicis proprius (PIN [radial nerve]: C7, C8)
  - EDM (PIN [radial nerve]: C7, C8)
- **Finger abduction ("DAB"):**
  - "DAB" = **D**orsal interossei **AB**duct
  - Four dorsal interossei (ulnar nerve: C8, T1)
  - Abductor digiti minimi (ulnar nerve: C8, T1)
- **Finger adduction ("PAD"):**
  - "PAD" = **P**almar interossei **AD**duct
  - Three palmar interossei (ulnar nerve: C8, T1)
- **Thumb flexors:**
  - Flexor pollicis brevis (superficial head: median nerve; deep head: ulnar nerve: C8, T1)
  - Flexor pollicis longus (AIN [median nerve]: C8, T1)
  - Opponens pollicis (median nerve: C8, T1)
  - Adductor pollicis (ulnar nerve: C8, T1)
- **Thumb extensors:**
  - EPL (PIN [radial nerve]: C7, C8)
  - EPB (PIN [radial nerve]: C7, C8)
  - APL (PIN [radial nerve]: C7, C8)
- **Thumb abduction:**
  - APL (PIN [radial nerve]: C7, C8)
  - Abductor pollicis brevis (median nerve: C8, T1)

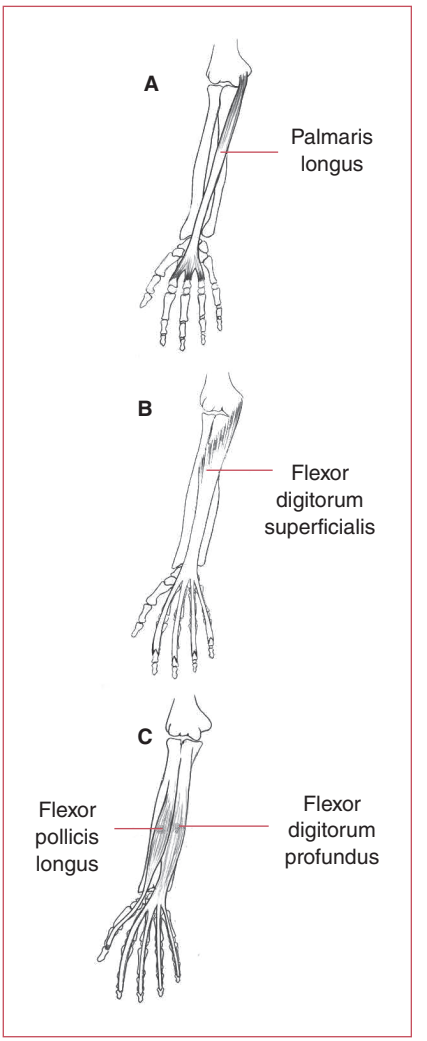

FIGURE 4–64 Hand flexors: Muscles of the forearm anterior compartment. (A) First layer; (B) second layer; (C) third layer.

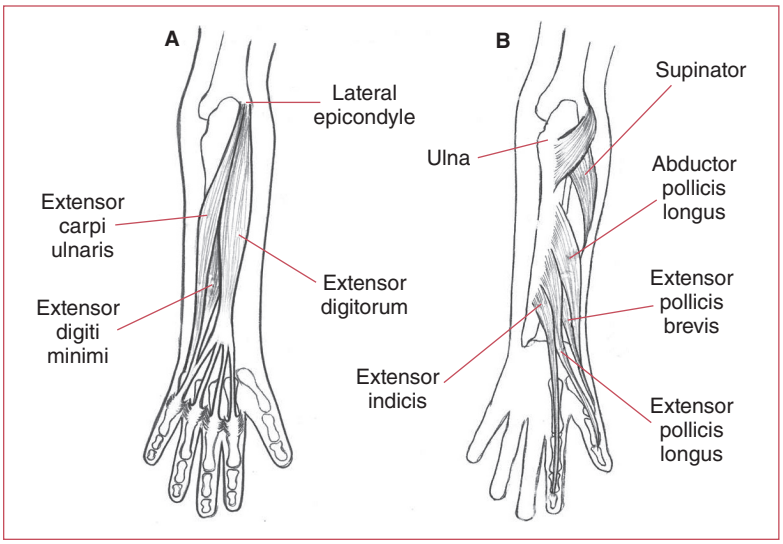

FIGURE 4–65 Muscle of the forearm posterior compartment. (A) Superficial layer; (B) deep layer.

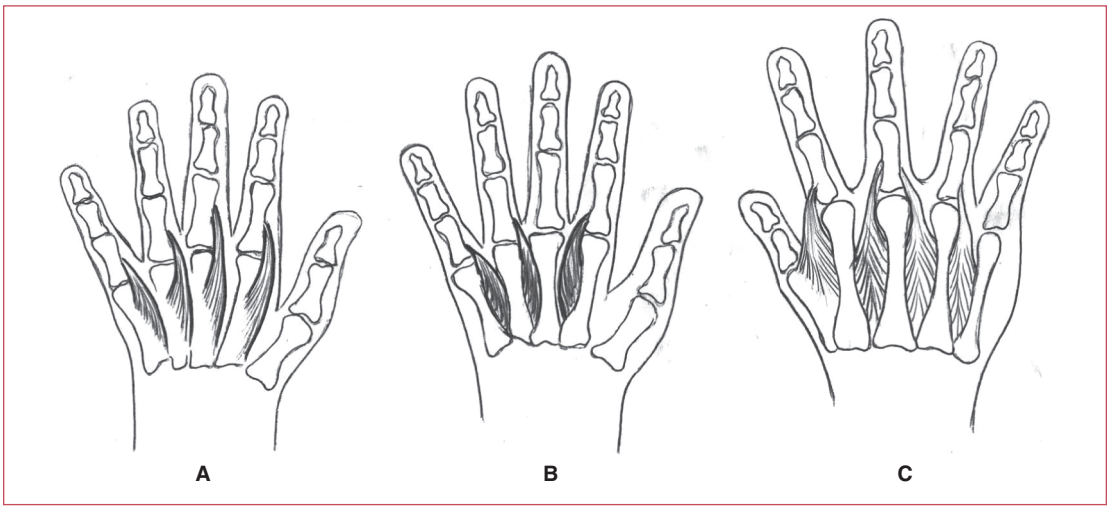

FIGURE 4–66 Palmar view: Intrinsic muscles of the hand. (A) Lumbricals; (B) palmar interossei; (C) dorsal interossei.

- **Thumb adduction:**
  - Adductor pollicis (ulnar nerve: C8, T1)
- **Opposition of the thumb to fifth digit:**
  - Opponens pollicis (median nerve: C8, T1)
  - Flexor pollicis brevis, superficial head (median nerve: C8, T1)
  - Abductor pollicis brevis (median nerve: C8, T1)
  - Opponens digiti minimi (ulnar nerve: C8, T1)

HAND DISORDERS

DUPUYTREN'S CONTRACTURE (FIGURE 4–67)

General

- Thickening and contraction of the palmar fascia due to fibrous proliferation

Etiology

- Unknown cause
- Has a genetic component primarily in people of Northern European descent. Previously known as the Viking disease.
- Commonly associated with DM, alcoholism, epileptics, and pulmonary tuberculosis (TB)
- Typically seen in men >40 years old

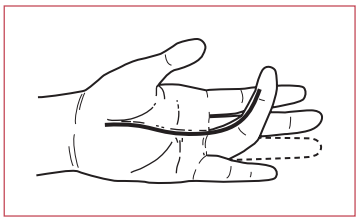

FIGURE 4–67 Dupuytren's contracture.

Clinical Features

- Painless nodules in the distal palmar crease. These nodules are initially nontender and may become tender as the disease progresses.
- The involved finger is drawn into flexion as the nodules thicken and contract.
- Flexion is commonly seen at the MCP joint involving the ring finger (fourth digit).

Imaging

- None needed

Treatment

- *Conservative:* Physical therapy, corticosteroid injections, collagenase injections. US, splinting, massage.
- *Surgical:* Surgical release if severe and affects function

STENOSING TENOSYNOVITIS: TRIGGER FINGER (FIGURE 4–68)

General

- Repetitive trauma that causes an inflammatory process to the flexor tendon sheath of the digits
- This process forms a nodule in the tendon, resulting in abnormal gliding through the A1 pulley system. As the digit flexes, the nodule passes under the pulley system and gets caught on the narrow annular sheath; as a result, the finger is locked in a flexed position.

ETIOLOGY

- Commonly associated with repetitive trauma, DM, RA, gout
- Seen in persons >40 years old

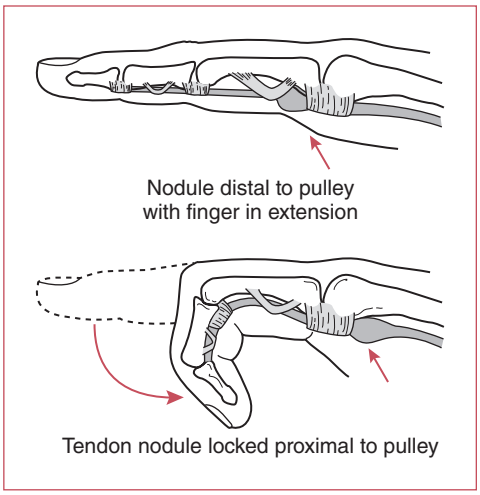

FIGURE 4–68 Trigger finger. Nodule or thickening in flexor tendon, which strikes the proximal pulley, making finger extension difficult.

Clinical Features

- A painful catching or locking with finger flexion and/or extension
- Palpable nodule may be tender on exam.

Imaging

- None needed

Treatment

- *Conservative:* Corticosteroid injection, immobilization by splinting, NSAIDs
- *Surgical*: Surgical release if conservative treatment fails

LIGAMENTOUS INJURIES (FIGURE 4–69)

General

- Involve the ligaments of the digits (PIP and MCP) and/or the thumb (MCP)
  - Ligaments: Collaterals and volar plate
- Injury may result in a partial tear (sprain) or complete dislocation.

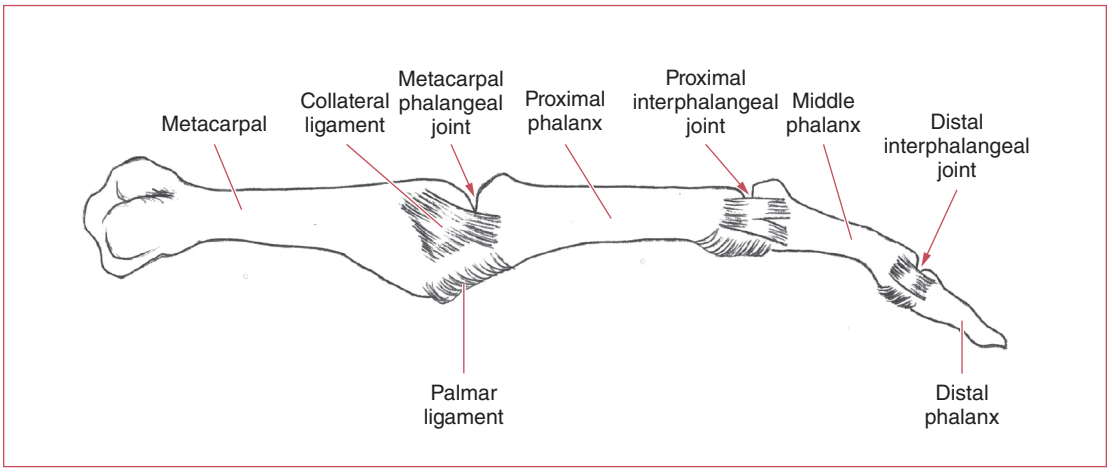

FIGURE 4–69 Ligaments of the MCP, PIP, and DIP (lateral view).
DIP, distal interphalangeal; MCP, metacarpophalangeal; PIP, proximal interphalangeal.

MECHANISMS OF INJURY

- MCP and PIP ligamentous injury to the digits and/or thumb (MCP)
  - Collateral ligament: Valgus or varus stress with the finger in an extended position
  - Volar plate: Hyperextension with dorsal dislocation, which is usually reducible
- MCP ligamentous injury to the thumb
  - UCL:
    - Test by placing valgus stress at the MCP joint of the thumb.
    - Also known as gamekeeper's thumb or skier's thumb (please refer to later section)
  - Radial collateral ligament: Uncommon

Clinical Features

- History of trauma to the finger with an immediate obvious deformity
- Local tenderness over the involved area with swelling of the joint
- Palpate both sides and assess the stability of the joint by applying a stress to the medial and lateral aspect.

Imaging

- AP and lateral views to rule out fracture and ensure proper reduction and congruency of the joint

Treatment

- *Conservative:* Simple dislocations
  - Reduce the joint by stabilizing the proximal end and applying a distal traction.
  - Buddy splinting of the finger should be done for approximately 2 weeks.
  - Thumb spica 3 to 6 weeks for MCP injuries.
- *Surgical:* Complex lesions

SKIER'S THUMB OR GAMEKEEPER'S THUMB

General

- Most often seen in skiers, basketball players, and other ball-handling athletes
- May occur with chronic lateral laxity or acute disruption of the UCL
- UCL attaches dorsally at the metacarpal head and runs distally to insert on the volar side of the proximal phalanx base. >80% of acute tears occur at the distal insertion point.
- Mechanism of injury is a forceful radial deviation of the proximal phalanx at the MCP joint often times with the thumb in an exposed abducted/extended position out of plane with the palm. Sudden or chronic hyperextension and/or hyperabduction at this joint can lead to partial or complete tear.
- Complete tears can lead to entrapment of the adductor aponeurosis between the ruptured portions of the ligament. This is referred to as Stener's lesion and will impair healing. Avulsion or avulsion fracture can also occur; both are surgical indications.

Clinical Features

- Instability of the MCP joint
- Prior to performing stress examination of the joint, x-ray should be obtained to rule out nondisplaced avulsion fracture.
- To examine, stabilize the radial portion of the MCP and position the joint in approximately 30 degrees of flexion. A radial deviation force is then applied distally to stress the UCL.
- Palpation of torn ligaments may identify Stener's lesion, which is felt as fullness at the MCP.
  - Grade I injury: Pain and no increased motion
  - Grade II injury: Increased opening with pain on stressing
  - Grade III injury: No pain, continued motion while stressing

Imaging

- Stress radiograph should be done comparing both hands. Instability indicated by radial deviation >40 degrees in extension and >20 degrees in flexion on plain films.
- Surgical referral should be considered above these thresholds of deviation.
- If radiographs are equivocal, one can obtain MRI or use US for dynamic evaluation.

Treatment

- Short arm cast with thumb spica splint for 4 to 6 weeks. Return to play or full activity when thumb is painless, with firm endpoint on radial deviation stress and at least 80% recovery of ROM and pinch strength.
- Stener's lesion with failure to heal, ligamentous avulsion, or avulsion fractures will likely require surgical treatment.

JERSEY FINGER (FIGURE 4–70)

General

- Complete or incomplete injury to the FDP tendon. Most commonly involves the fourth digit.
- Most commonly due to trauma as seen in athletes (football, wrestling). May also be spontaneous (as in the case of RA).
- The classic mechanism of injury in athletes is when a player's finger gets caught in the jersey of another when attempting to grab them. The forceful DIP extension while the FDP muscle is contracting can result in injury to the profundus tendon.

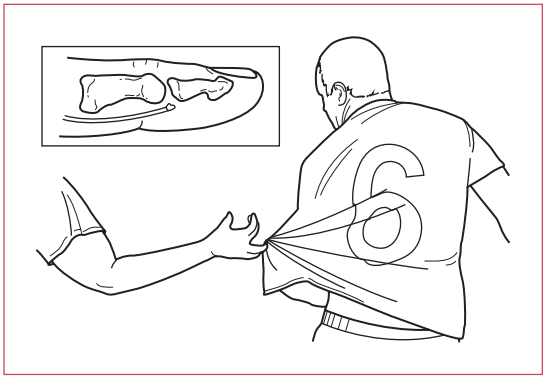

FIGURE 4–70 Jersey finger: Mechanism of injury is rupture of the profundus tendon.

The profundus tendon can be avulsed from its insertion and possibly accompanied by an avulsion fracture.

Clinical Features

- The patient is unable to actively flex the DIP joint.
- Testing of the FDP (**Figure 4–71A**):
  - Flex the DIP while the PIP joint is held in extension. The action of the FDS is eliminated when the PIP is maintained in extension.
- Testing flexion of the FDS (**Figure 4–71B**):
  - It is important to eliminate the action of the FDP because the FDP can perform many of the same actions as the FDS (MCP and PIP flexion) secondary to its distal attachment at the DIP.
  - Hold the DIP of the noninvolved digits in extension. Then ask the patient to flex the unrestrained digit, which can only be done with a normal FDS tendon. This maneuver isolates the FDS and eliminates action of the FDP.

Imaging

- X-ray films may show an avulsed fragment near tendinous insertion.

Treatment

- *Conservative:* Little regained by conservative care
- *Surgical:* Early orthopedic referral for surgical repair

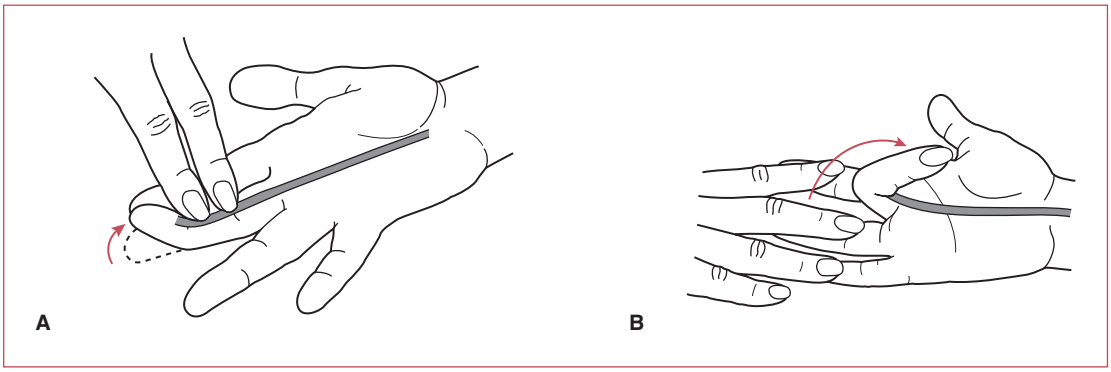

FIGURE 4–71 (A) Test for FDP function. (B) Test for FDS function. FDP, flexor digitorum profundus; FDS, flexor digitorum superficialis.

MALLET FINGER (FIGURE 4–72)

General

- Commonly known as baseball finger
- Sudden passive flexion of the DIP joint when the finger is extended, causing a rupture of the extensor tendon
- An avulsion fracture of the distal phalanx may also occur.

Clinical Features

- A flexed DIP joint that cannot be actively extended
- DIP joint tenderness and edema at the distal dorsal area

Imaging

- X-ray of the hand to evaluate for an avulsion fracture of the distal phalanx

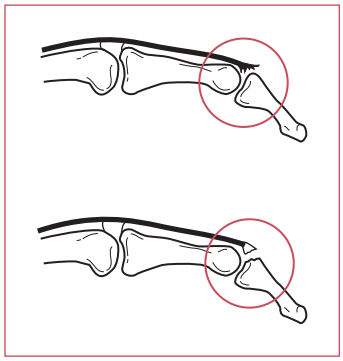

FIGURE 4–72 Mallet finger. Top: Rupture of the extensor tendon at its insertion. Bottom: Avulsion of a piece of distal phalanx.

Treatment

- *Conservative:* Splinting of the DIP in extension for 6 to 8 weeks **(Figure 4–73)** with a stack splint or custom-made splint.
  - Maintaining the DIP in extension at all times is essential.
  - Weekly visits to assess full finger flexion should be done.
  - At the end of the 6-week course, gentle active flexion with night splinting should be done for 2 to 4 weeks.
- *Surgical:*
  - Surgical repair reserved for poor healing or if an avulsed fragment involves greater than one-third of the joint.

FIGURE 4–73 Stack splint for treatment of mallet finger.

FRACTURE OF THE BASE OF THE FIRST METACARPAL

BENNETT'S AND ROLANDO'S FRACTURE

General

- **Bennett's fracture** (Figure 4–74A): Oblique fracture-subluxation at the base of the thumb metacarpal
- **Rolando's fracture** (Figure 4–74B) Fracture at the base of the thumb metacarpal that may be classified as a T, Y, or comminuted configuration

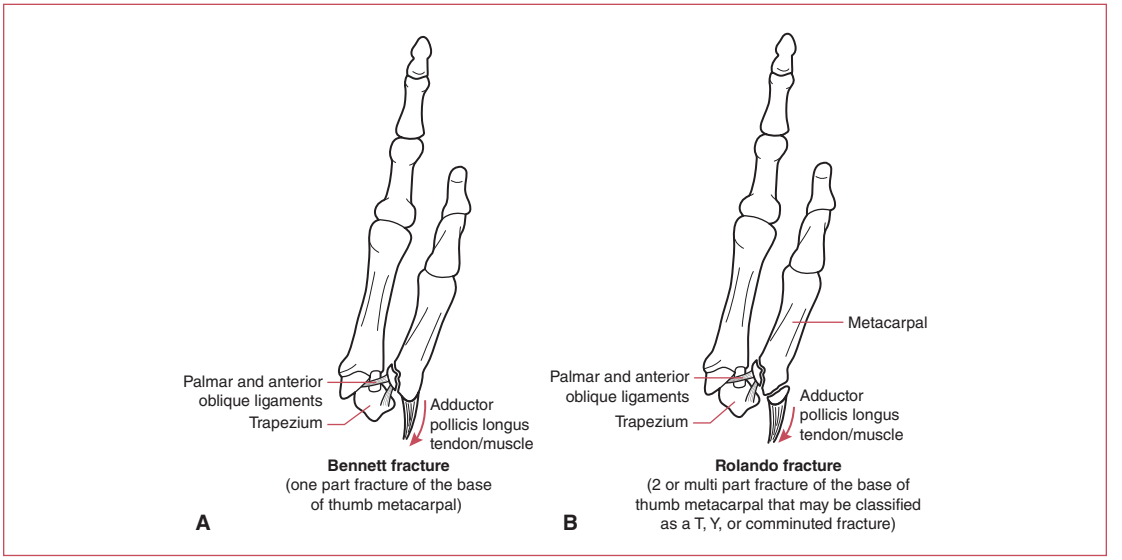

FIGURE 4–74 Bennett's (A) and Rolando's (B) fractures.

COMPLICATIONS

- An avulsed metacarpal fragment in a Bennett's fracture may sublux secondary to the proximal pull of the APL muscle.

Clinical Features

- Tenderness and swelling at the base of the digit (thumb or fifth digit) following a direct blow to a flexed thumb or digit

Imaging

- X-ray: AP, lateral, and oblique views

Treatment

- Orthopedic referral

METACARPAL NECK OR SHAFT FRACTURE (FIGURE 4–75)

General

- Also known as a Boxer's fracture
- Fracture of the metacarpal neck/shaft usually seen after a person strikes a wall or another person
- May occur at any digit but commonly seen in the fifth digit

Clinical Features

- Tenderness and swelling in the area of the hand seen after the traumatic event

Imaging

- X-rays

Treatment

- Orthopedic referral

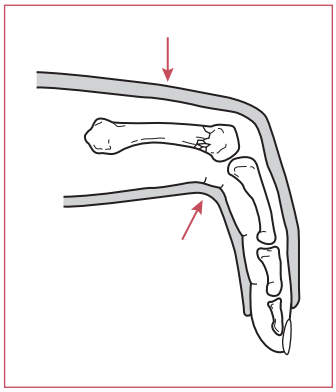

FIGURE 4–75 Boxer's fracture (placed in an ulnar gutter splint).

LOWER EXTREMITIES: THE HIP AND PELVIS

- The five joints of the pelvic girdle include the bilateral femoroacetabular (hip) joints, the pubic symphysis, and the bilateral sacroiliac (SI) joints.
- The hip is a very stable, multidirectional mobile ball-and-socket joint (enarthrosis).
- Due to high mobility, hip joint pathology will be manifested during weight bearing, ambulation, or motion.
- Pathology affecting the SI joint and pubic symphysis does not restrict motion to the extent that hip joint pathology will.
- The angle between the femoral neck and shaft of the femur is different between males (125 degrees) and females (115–120 degrees). This difference is due to the female pelvis being wider to accommodate the birth canal and gravid uterus.
  - Coxa vara occurs when the femoral neck and shaft angle is decreased. The affected leg is shortened, and hip abduction is limited. The knee assumes a valgus deformity.
  - Coxa valga occurs when the angle is increased. The affected limb is lengthened, and the knees assume a varus deformity.

HIP AND PELVIC FUNCTIONAL ANATOMY (FIGURE 4–76)

Muscles

HIP FLEXORS (FIGURE 4–77)

HIP FLEXORS	SPINAL INNERVATION	PERIPHERAL INNERVATION
Iliopsoas (primary hip flexor)	L1, L2, L3, L4	Direct branches of L1–L4 anterior rami
Sartorius	L2, L3, L4	Femoral nerve
Rectus femoris	L2, L3, L4	Femoral nerve

(*continued*)

HIP FLEXORS	SPINAL INNERVATION	PERIPHERAL INNERVATION
Pectineus	L2, L3, L4	Femoral nerve
Adductor brevis	L2, L3, L4	Obturator nerve
Adductor longus	L2, L3, L4	Obturator nerve
Gracilis	L2, L3, L4	Obturator nerve
Adductor magnus	L2, L3, L4, L5, S1	Obturator and sciatic (tibial division) nerves
Tensor fasciae lata (TFL)	L4, L5, S1	Superior gluteal nerve

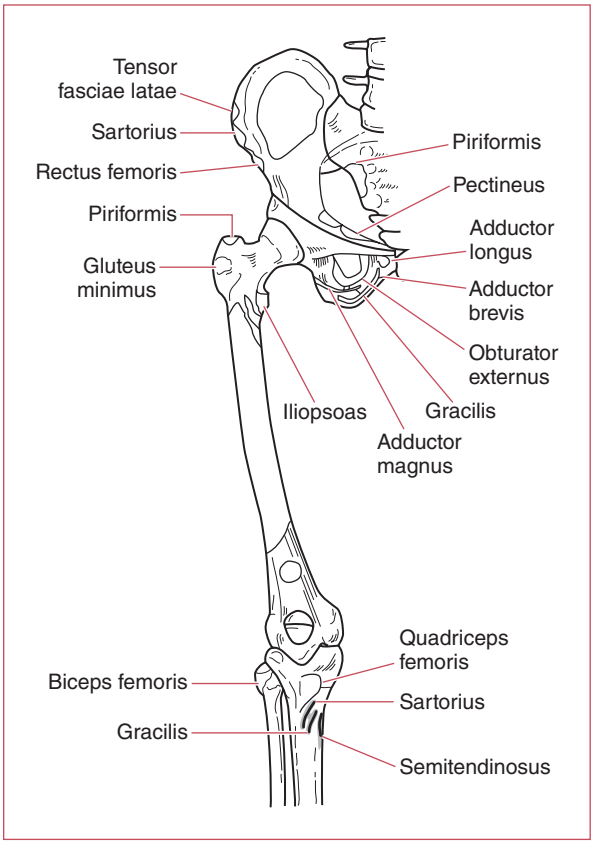

FIGURE 4–76 The pelvis, thigh, and knee region.

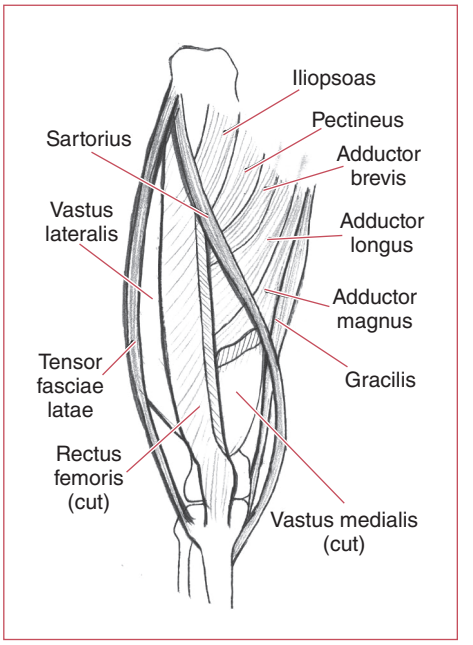

FIGURE 4–77 Hip flexors (anterior view).

Hip Adductors (Anteriorly Placed; Figure 4–78)

ANTERIOR HIP ADDUCTORS	SPINAL INNERVATION	PERIPHERAL INNERVATION
Gracilis	L2, L3, L4	Obturator nerve
Adductor brevis	L2, L3, L4	Obturator nerve
Adductor longus	L2, L3, L4	Obturator nerve
Pectineus	L2, L3, L4	Femoral nerve
Adductor magnus	L2, L3, L4, L5, S1	Obturator nerve and sciatic (tibial division) nerves

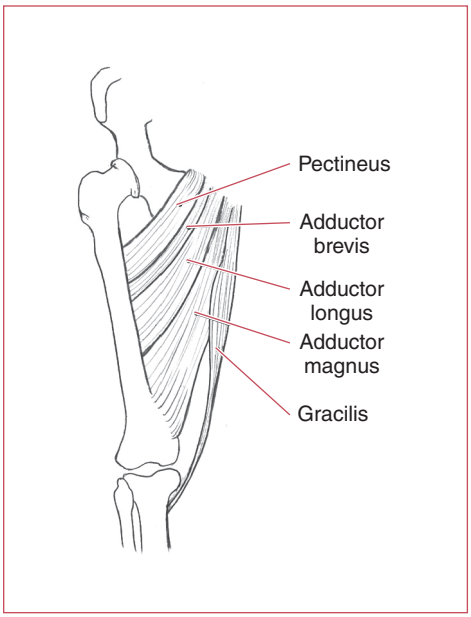

FIGURE 4–78 Adductors of the hip (anterior view).

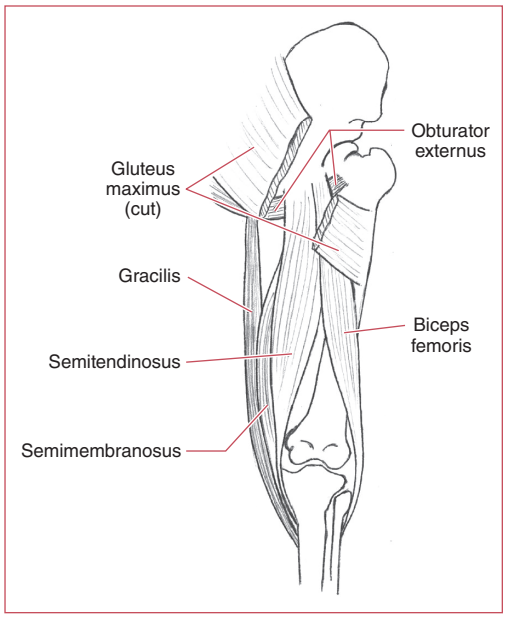

FIGURE 4–79 Adductors of the hip (posterior view).

Hip Adductors (Posteriorly Placed; Figure 4–79)

POSTERIOR HIP ADDUCTORS	SPINAL INNERVATION	PERIPHERAL INNERVATION
Gracilis	L2, L3, L4	Obturator nerve
Obturator externus	L3, L4	Obturator nerve
Gluteus maximus	L5, S1, S2	Inferior gluteal nerve
Long head biceps femoris	L5, S1, S2	Sciatic nerve (tibial division)
Semimembranosus	L5, S1, S2	Sciatic nerve (tibial division)
Semitendinosus	L4, L5, S1, S2	Sciatic nerve (tibial division)

Hip Abductors

HIP ABDUCTORS	SPINAL INNERVATION	PERIPHERAL INNERVATION
Gluteus medius Gluteus minimus	L4, L5, S1	Superior gluteal nerve

Hip Abductors and Internal Rotators

HIP ABDUCTORS AND INTERNAL ROTATORS	SPINAL INNERVATION	PERIPHERAL INNERVATION
Sartorius	L2, L3, L4	Femoral nerve
Tensor fasciae lata	L4, L5, S1	Superior gluteal nerve
Gluteus maximus, superior fibers	L5, S1, S2	Inferior gluteal nerve
Piriformis	L5, S1, S2	Nerve to piriformis

Hip Extensors (Figure 4–80)

HIP EXTENSORS	SPINAL INNERVATION	PERIPHERAL INNERVATION
Gluteus maximus *Primary hip extensor	L5, S1, S2	Inferior gluteal nerve
Gluteus medius, posterior fibers Gluteus minimus, posterior fibers	L4, L5, S1	Superior gluteal nerve
Piriformis	S1, S2	Nerve to piriformis
Adductor magnus	L2, L3, L4	Sciatic nerve (tibial division) for extension; obturator nerve for adduction
Hamstring muscles – Long head of the biceps femoris – Semimembranosus – Semitendinosus	 L5, S1, S2 L5, S1, S2 L5, S1, S2	Sciatic nerve (tibial division)

External (Lateral) Rotators of the Hip (Figure 4–81)

HIP EXTERNAL ROTATORS	SPINAL INNERVATION	PERIPHERAL INNERVATION
Gluteus maximus	L5, S1, S2	Inferior gluteal nerve
Piriformis	S1, S2	Nerve to piriformis
Obturator internus	L5, S1	Nerve to the obturator internus
Obturator externus	L5, S1, S2	Obturator nerve
Superior gemellus	L5, S1, S2	Nerve to the superior gemellus
Inferior gemellus	L5, S1, S2	Nerve to the inferior gemellus
Quadratus femoris	L4, L5, S1	Nerve to quadratus femoris

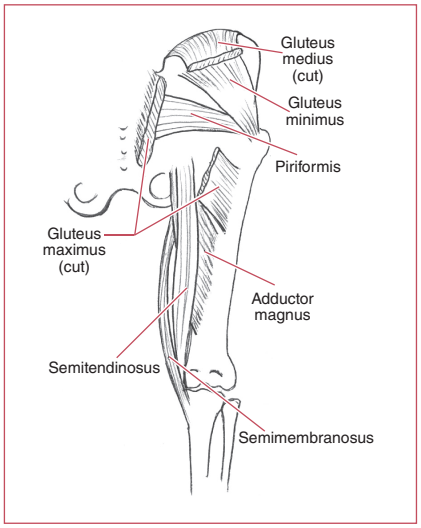

FIGURE 4–80 Extensors of the hip (posterior view).

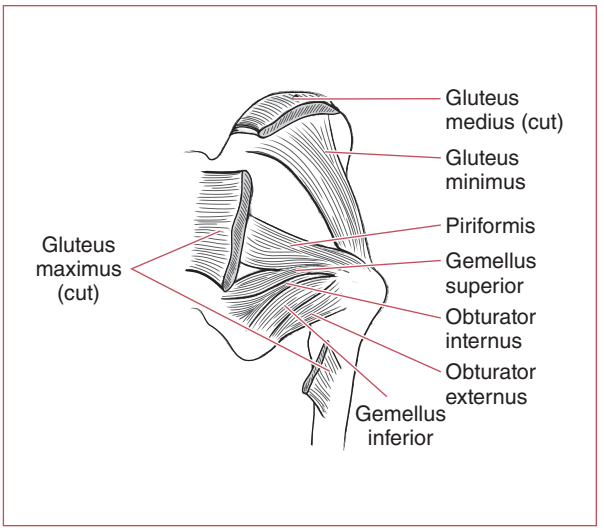

FIGURE 4–81 External (lateral) rotators of the thigh (quadratus femoris not shown; posterior view).

Internal Rotators of the Hip (Figure 4–82)

INTERNAL (MEDIAL) ROTATION

- Mnemonic: TAGGGSS

HIP INTERNAL ROTATORS	SPINAL INNERVATION	PERIPHERAL INNERVATION
Tensor fasciae lata	L4, L5, S1	Superior gluteal nerve
Adductor magnus	L2, L3, L4, L5, S1	Obturator nerve and sciatic nerve (tibial division)
Adductor longus and brevis	L2, L3, L4	Obturator nerve
Gracilis	L2, L3, L4	Obturator nerve
Gluteus medius, posterior fibers Gluteus minimus, posterior fibers	L4, L5, S1	Superior gluteal nerve
Semitendinosus	L5, S1, S2	Sciatic nerve (tibial division)
Semimembranosus	L5, S1, S2	Sciatic nerve (tibial division)

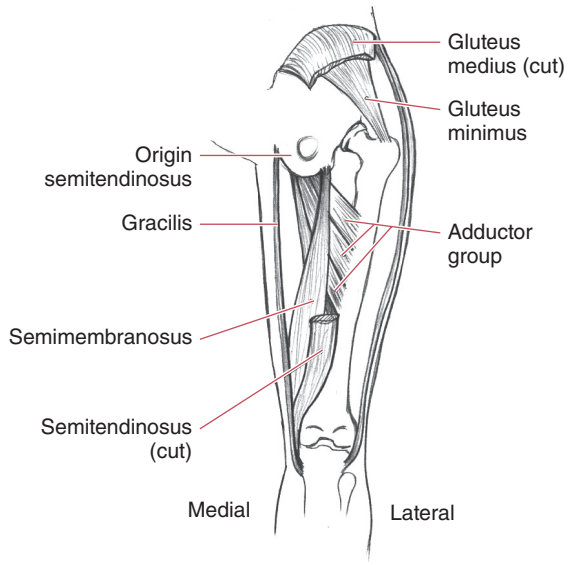

FIGURE 4–82 Internal (Medial) rotators of the thigh (posterior view).

Ligaments (Figure 4–83A and B)

ACETABULAR LABRUM (GLENOID LABRUM)

- The acetabular labrum serves to deepen the acetabulum. Its function is to hold the femoral head in place.

ARTICULAR CAPSULE

- The fibrous articular capsule extends from the acetabular rim to the intertrochanteric crest, forming a cylindrical sleeve that encloses the hip joint and most of the femoral neck. Circular fibers around the femoral neck constrict the capsule and help to hold the femoral head in the acetabulum.

ILIOFEMORAL LIGAMENT

- Also known as the Y-ligament of Bigelow, it is the strongest ligament in the body.
- The iliofemoral ligament extends from the anterior inferior iliac spine (AIIS) to the intertrochanteric line.
- Its function is to limit extension, abduction, and external rotation of the hip.

ISCHIOFEMORAL LIGAMENT

- The ischiofemoral ligament extends from the ischium behind the acetabulum to blend with the capsule.
- Its function is to limit internal rotation of the hip.

PUBOFEMORAL LIGAMENT

- The pubofemoral ligament extends from the superior pubic ramus and joins the iliofemoral ligament.
- Its function is to limit hip abduction.

LIGAMENTUM CAPITIS FEMORIS

- The capitis femoris ligament extends from the acetabular notch to the femur.
- This ligament is weak and does little to strengthen the hip.
- In 80% of cases, it carries a small artery to the femoral head.

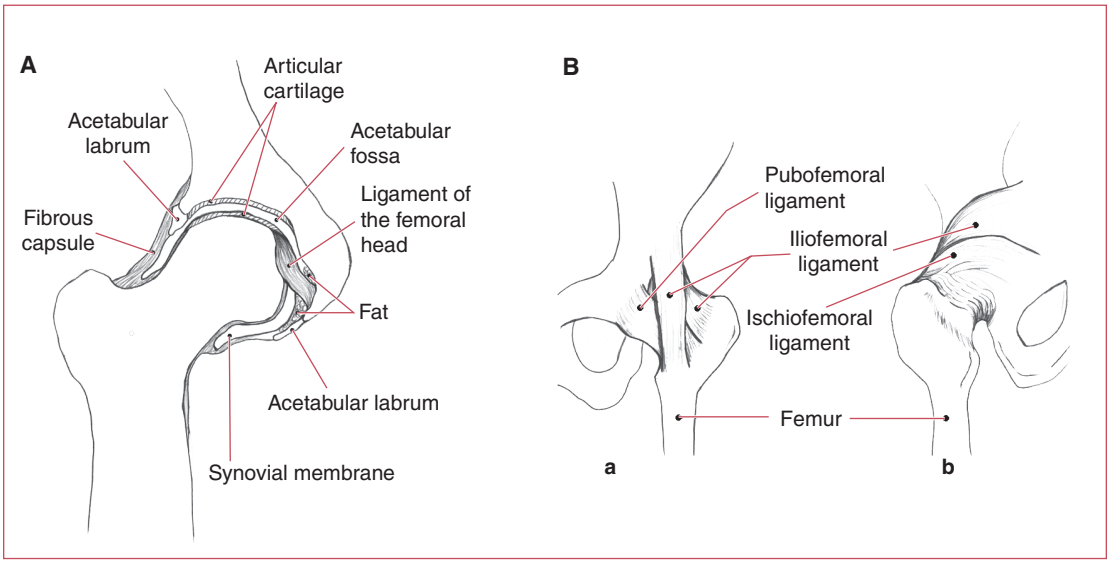

FIGURE 4–83 (A) Frontal section through the hip joint. (B) Anterior (a), and posterior (b) view of the left hip joint.

NORMAL RANGE OF HIP MOTION IN THE ADULT

ADULT HIP ROM	DEGREES
Hip flexion	110–130 degrees
Hip extension	30 degrees
Hip abduction	45–50 degrees
Hip adduction	0–30 degrees
Hip external rotation	35 degrees (ER hip is usually greater than IR)
Hip internal rotation	45 degrees

ER, external rotation; IR, internal rotation; ROM, range of motion.

- 🕮 OA will limit internal rotation of the hip first.

HIP TESTS

FABERE (Patrick's) Test (Figure 4–84)

- Provocative maneuver to assess for intra-articular hip pathology or SI joint dysfunction
- Motions of the test: **F**lexion, **AB**duction, **E**xternal **R**otation, and **E**xtension (FABERE)
- With the patient supine, passively flex, abduct the hip, and externally rotate. Extension of the leg is achieved with a downward force by the examiner.
  - Anterior hip/groin pain is indicative of intra-articular or periarticular hip pathology.
  - Posterior hip pain is indicative of an SI joint disorder.

FIGURE 4–84 FABERE (Patrick's) test. FABERE, Flexion, ABduction, External Rotation, and Extension.
Source: Courtesy of JFK Johnson Rehabilitation Institute, 2000.

🕮 Thomas Test (Figure 4–85)

- This test is used to assess hip flexion contractures.
- Perform this test with the patient supine: Flex one hip, fully reducing the lumbar spine lordosis. Stabilize the lumbar spine/pelvis and extend the contralateral hip. If that hip does not fully extend, a flexion contracture is present.

Ober Test (Figure 4–86)

- Tests for contraction of the tensor fascia lata (TFL)/iliotibial band (ITB) tightness
- With the patient side-lying with the uninvolved leg on the table, flex the knee to 90 degrees, extend the hip to 0 degrees, and abduct the involved leg as far as possible. The leg is then lowered from full abduction.
- If the thigh remains abducted, there may be a contracture of the TFL or ITB.

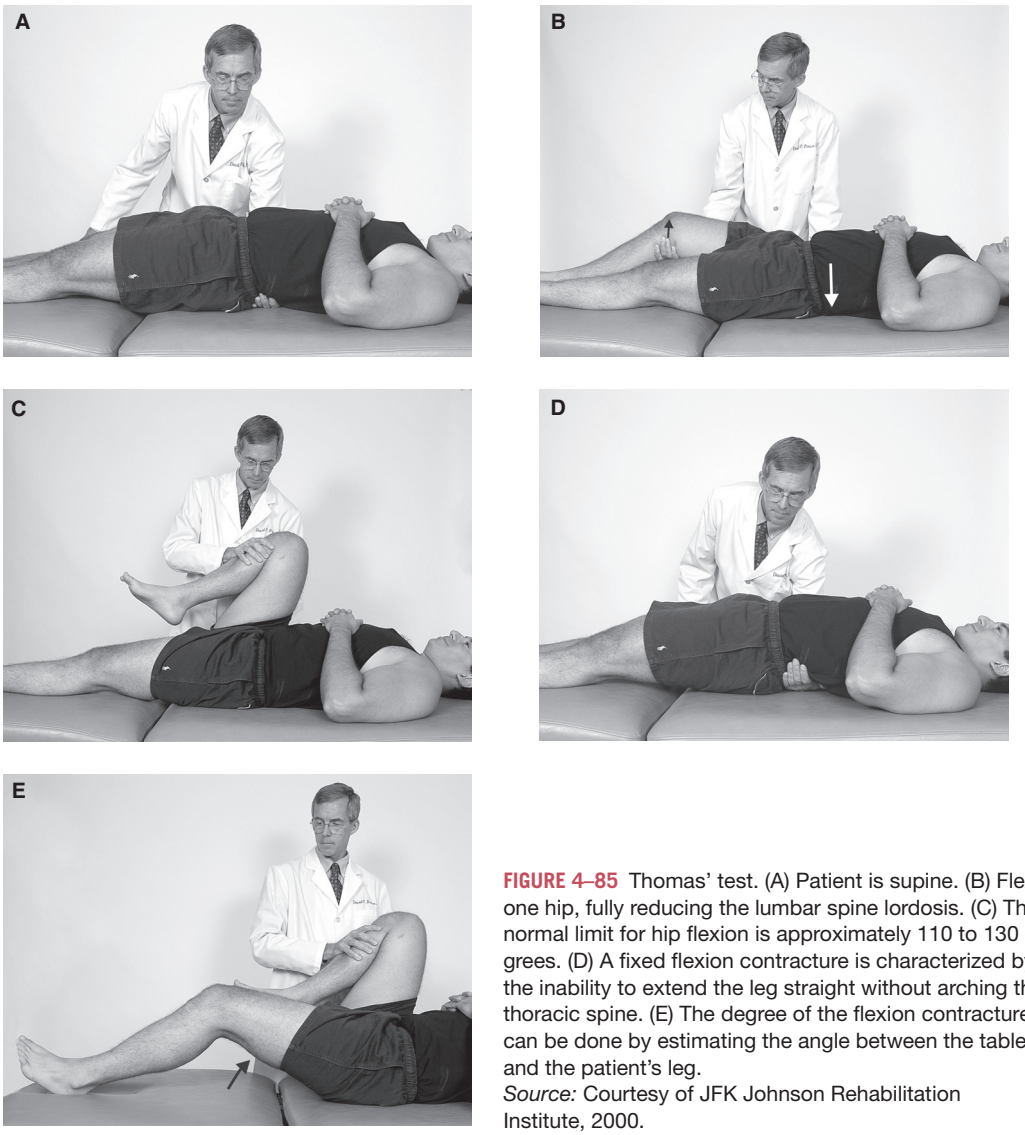

FIGURE 4–85 Thomas' test. (A) Patient is supine. (B) Flex one hip, fully reducing the lumbar spine lordosis. (C) The normal limit for hip flexion is approximately 110 to 130 degrees. (D) A fixed flexion contracture is characterized by the inability to extend the leg straight without arching the thoracic spine. (E) The degree of the flexion contracture can be done by estimating the angle between the table and the patient's leg.
Source: Courtesy of JFK Johnson Rehabilitation Institute, 2000.

Trendelenburg Test (Figure 4–87)

- Tests for gluteus medius weakness
- With the patient standing, ask them to raise one foot off the ground.
- Strength of the gluteus medius on the supported side is assessed.
  - A positive test occurs when the pelvis on the unsupported side descends. Example: Pelvic drop on the left side in a patient standing on their right leg is indicative of right gluteus medius weakness.
  - A negative test occurs when the pelvis on the unsupported side stays the same height or elevates slightly.
- Conditions associated with gluteus medius weakness:
  - Radiculopathies
  - Poliomyelitis
  - Meningomyelocele
  - Fractures of the greater trochanter
  - Slipped capital femoral epiphysis (SCFE)
  - Congenital hip dislocation
  - Deconditioning

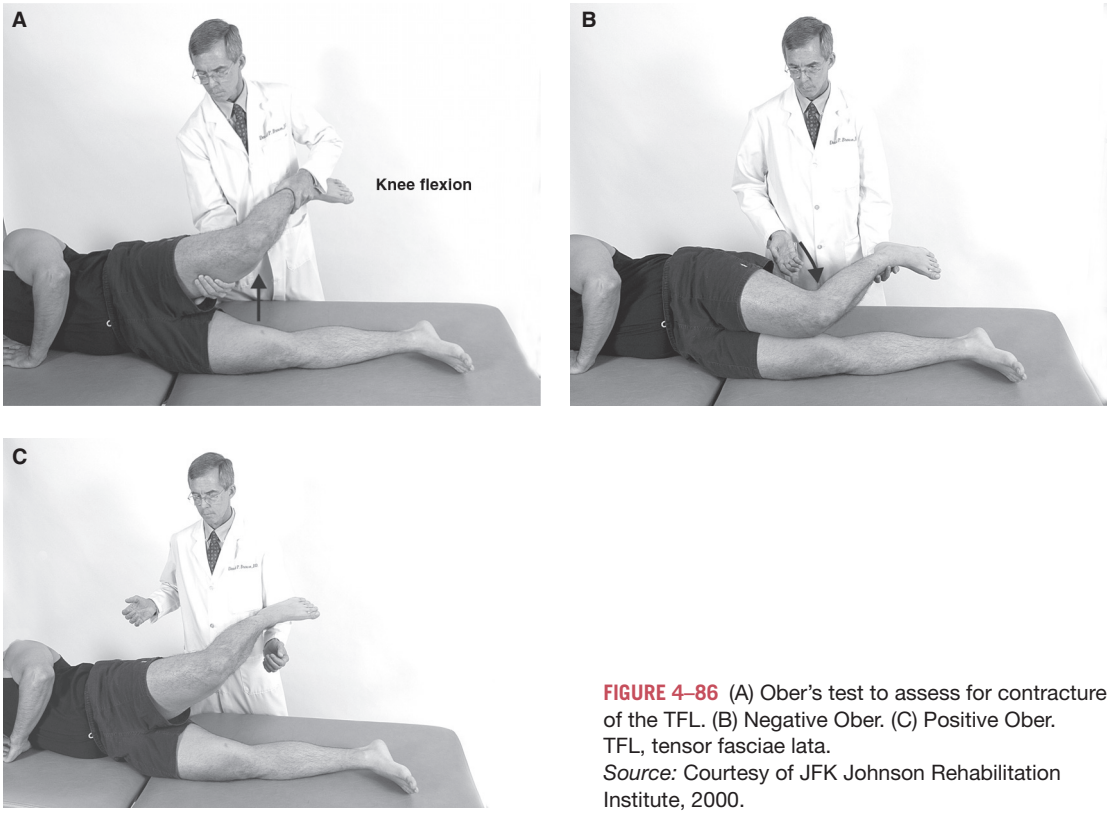

FIGURE 4–86 (A) Ober's test to assess for contracture of the TFL. (B) Negative Ober. (C) Positive Ober. TFL, tensor fasciae lata.
Source: Courtesy of JFK Johnson Rehabilitation Institute, 2000.

FIGURE 4–87 Trendelenburg test. (A) Negative. (B) Positive.
Source: Courtesy of JFK Johnson Rehabilitation Institute, 2000.

Femoral Nerve Stretch Test (Ely's Test)

- Tests for femoral nerve irritation
- With the patient lying prone, flex the knee >90 degrees and extend the hip.
- Pain in the anterior thigh is positive for femoral nerve irritation.

LEG LENGTH DISCREPANCY

True Leg Length Discrepancy (Figure 4–88)

- 🕮 To assess true leg length, measure from the anterior superior iliac spine (ASIS) to the medial malleolus.
  - Note that these are two fixed bony landmarks.

- To determine if the discrepancy is in the femur or the tibia:
  - With the patient supine, flex the knees 90 degrees and place the feet flat on the table.
  - If one knee is higher than the other, that tibia is longer **(Figure 4–88C)**.
  - If one knee projects further anteriorly, then that femur is longer **(Figure 4–88D)**.
  - True leg length discrepancy has many causes, including fractures crossing the epiphyseal plate in childhood or poliomyelitis.

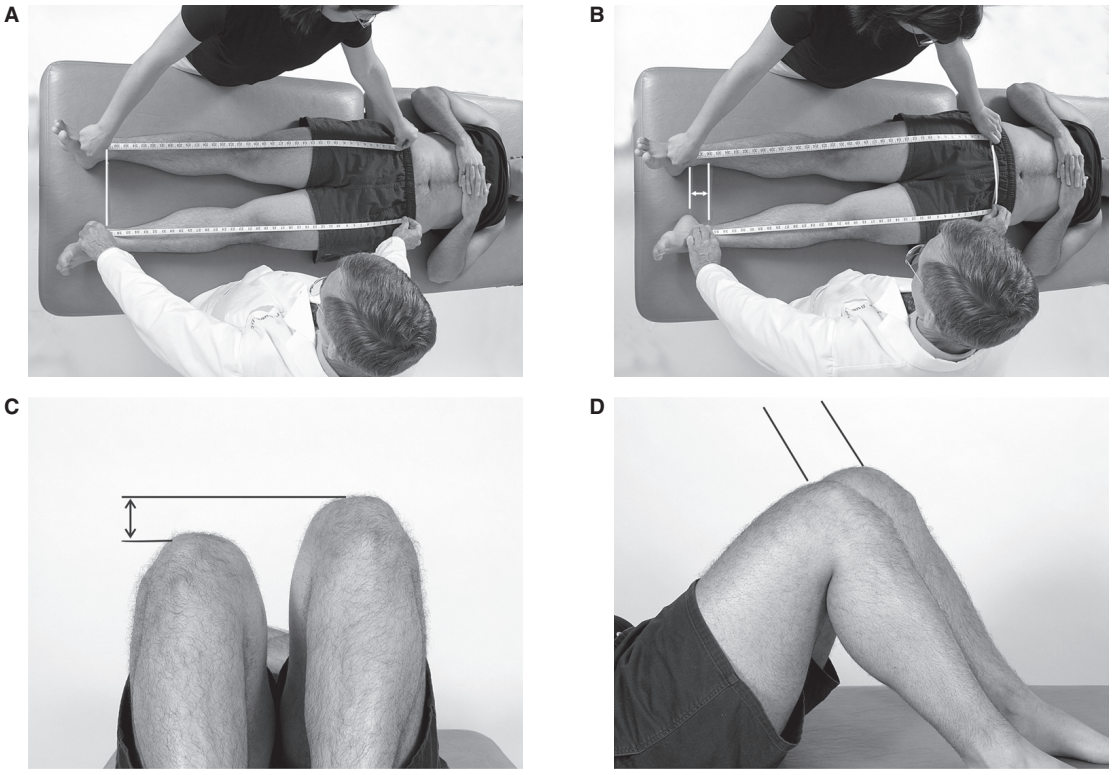

FIGURE 4–88 Leg length discrepancy. (A) Examiner should measure from one fixed bony point (i.e., ASIS to another (i.e., medial malleolus) to find true leg length. (B) True leg length discrepancy. (C) Tibial length discrepancy. (D) Femoral length discrepancy.
ASIS, anterior superior iliac spine.
Source: Courtesy of JFK Johnson Rehabilitation Institute, 2000.

Apparent Leg Length Discrepancy (Figure 4–89)

- First, determine that no true leg length discrepancy exists.
- Apparent leg length discrepancy may be caused by pelvic obliquities or flexion/adduction deformity of the hip.
- With the patient supine, measure from the umbilicus to the medial malleoli (from a nonfixed to a fixed landmark).
- Pelvis obliquity may be assessed by observing the levelness of the ASIS or the posterior superior iliac spines (PSIS).

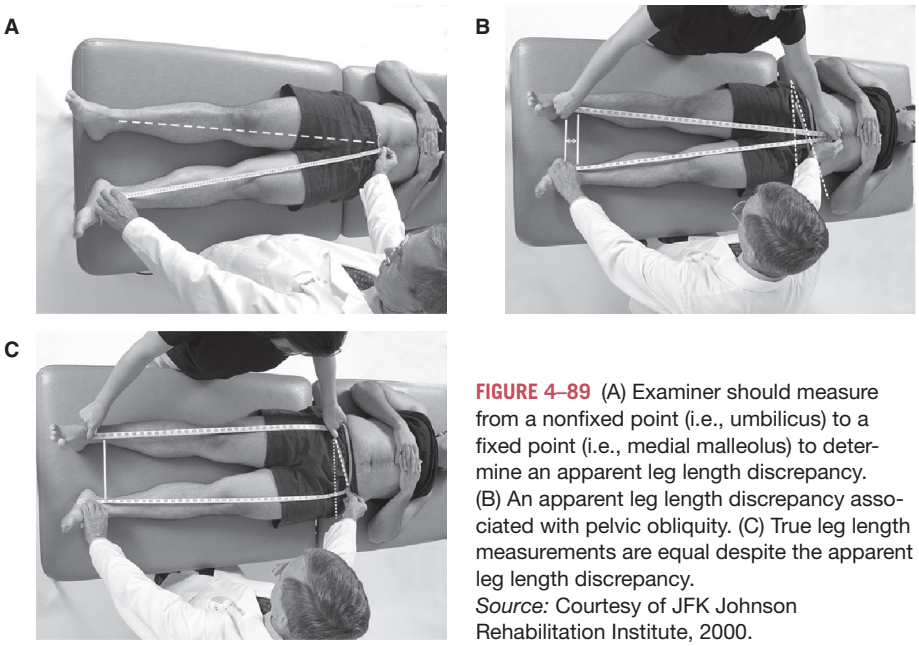

FIGURE 4–89 (A) Examiner should measure from a nonfixed point (i.e., umbilicus) to a fixed point (i.e., medial malleolus) to determine an apparent leg length discrepancy. (B) An apparent leg length discrepancy associated with pelvic obliquity. (C) True leg length measurements are equal despite the apparent leg length discrepancy.
Source: Courtesy of JFK Johnson Rehabilitation Institute, 2000.

HIP DISORDERS

HAMSTRING STRAIN

General

- Note that the normal strength ratio of hamstrings to quadriceps is 3:5.
- The hamstrings are placed under maximal stretch when the hip is forced into flexion and the knee into extension.
- Injuries typically occur during the eccentric phase of muscle contraction and at the myotendinous junction, most commonly in the lateral hamstrings.
- Predisposing factors associated with this strain include inadequate warm-up, poor flexibility, exercise fatigue, poor conditioning, and muscle imbalance.
  - A rehab program needs to correct these risk factors as well as core stability deficits.
- Injuries range in severity from Grade I (strain) to Grade III (complete tear).
- Most commonly seen in track and gymnastics injuries.

Clinical Features

- Presents as pain in the hamstring region after a forceful hamstring contraction or knee extension.
- Pain may occur with loss of function.
- There is tenderness over the muscle belly or origin.
- The examiner should attempt to stretch the injured muscle while palpating.
- Ecchymosis may descend to the thigh and present at the distal thigh or back of the knee or calf.

PROVOCATIVE TEST

- Pain elicited in the ischial region with knee flexion and tenderness to palpation

Imaging

- Generally not needed
- If warranted, plain films to look for avulsion fracture of the ischial tuberosity
- MRI can confirm diagnosis.

Treatment

- Ice, compression, activity restriction, NSAIDs
- Rehabilitation program:
  - Gentle stretch and ROM exercises
  - Advance to strengthening, gradually transitioning from concentric to eccentric exercises when tolerated
  - Core stabilization/strengthening, and risk factor modification when inflammation is reduced
- Injury prevention: Maintain hamstring flexibility and strength, with focus on eccentric exercises; core strengthening, neuromuscular control exercises, and sport-specific exercises.
- Return to play: Variable but typically ranges from 3 weeks to 6 months depending on the severity of injury.

HIP FLEXOR STRAIN

General

- Commonly seen in sprinting as well as in soccer, gymnastics, baseball, and football
- Occur due to eccentric overload of the psoas muscle or as the athlete tries to flex the fully extended hip, such as in hurdling or kicking

Clinical Features

- Tenderness to palpation over the area and with resisted hip flexion and passive hip extension

Imaging

- AP and frog leg lateral views are used to exclude bony injury such as an apophyseal avulsion fracture (commonly seen at ASIS, ischial tuberosity, AIIS, lesser trochanter, iliac crest).
- Injury to the apophyseal plate can occur in adolescent athletes.

Treatment

- Protected weight bearing, icing, and gentle AROM as soon as possible
- Strengthening exercises when gait is nonantalgic and ROM is full and pain-free
- Progress strength exercises from closed to open kinetic chain exercises and eccentric and plyometric training to prevent recurrent injury

PIRIFORMIS SYNDROME

General

- A painful muscle condition involving the piriformis muscle, an external hip rotator
- Piriformis syndrome can be stressed due to poor body mechanics in a chronic condition or an acute injury with forceful hip internal rotation.
- In severe spasms, the sciatic nerve may be involved to some degree because the nerve pierces the piriformis muscle fibers in some individuals.
- Rehabilitation seeks to reduce pain and spasm and recover full hip internal rotation.

Clinical Features

- Pain associated with piriformis injury may present in the lateral buttock, posterior hip, and proximal posterior thigh, as well as the SI region.
- The condition may be exacerbated by walking up stairs.
- There is tenderness over the muscle belly that stretches from the sacrum to the greater trochanter.

PROVOCATIVE TEST
- FAIR test
  - Pain with hip Flexion, Adduction, and Internal Rotation (FAIR)

Imaging
- Imaging the lumbar spine and hip may be necessary to rule out other etiologies or causes of pain.

Treatment
- Stretching of the external rotator hip muscles, NSAIDs, and US are the initial therapies.
- Corticosteroid injections can be used if more conservative measures fail.

ILIOPSOAS BURSITIS AND TENDONITIS

General
- Inflammation of the muscle tendon unit and bursa occurs with overuse or trauma, causing muscle tightness and imbalance.
- This condition may cause one type of snapping hip syndrome.

Clinical
- Hip snapping may occur with flexion and may cause pain.
- There is tenderness over the iliopsoas region.

PROVOCATIVE TEST
- Pain on resisted hip flexion

Imaging
- Radiographs of the hip are useful to rule out underlying bony pathology.

Treatment
- Ice, NSAIDs, stretching, and strengthening
- Corticosteroid injection if conservative measures fail
- Stretching the ITB, hip flexors, and gluteus maximus

SNAPPING HIP SYNDROME (ILIOTIBIAL BAND SYNDROME; FIGURE 4–90)

General
- Audible "snap" or click at the hip with ROM/ambulation
- Divided into internal and external snapping hip syndromes **(Figure 4–90)**
- **External snapping hip syndrome:**
  - May be due to a tight ITB or gluteus maximus snapping over the greater trochanter
- **Internal snapping hip syndrome:**
  - May be a result of a tight iliopsoas tendon/iliopsoas tendonitis snapping over the iliopectineal prominence of the pelvis
  - Less commonly, the patient may have an acetabular labral tear or loose body in the hip joint.

Clinical Features
- Patients may complain of hip snapping or clicking with or without pain.
- Tenderness over TFL/ITB or gluteus maximus with external snapping hip syndrome

- Tenderness in anterior groin (iliopsoas, labral tear, or loose body) with internal snapping hip syndrome. Patients may also have tenderness over anterior groin/inferior abdomen with iliopsoas tendonitis.

PROVOCATIVE TESTS

- External snapping hip syndrome: Internally and externally rotate the hip passively while the patient is in the lateral decubitus position.
- Internal snapping hip syndrome: Extend, abduct, and externally rotate the affected hip.

Imaging

- X-rays not needed

Treatment

- Relative rest, ice, and NSAIDs
- Rehab focuses on correction of biomechanics, as well as ROM/stretching.

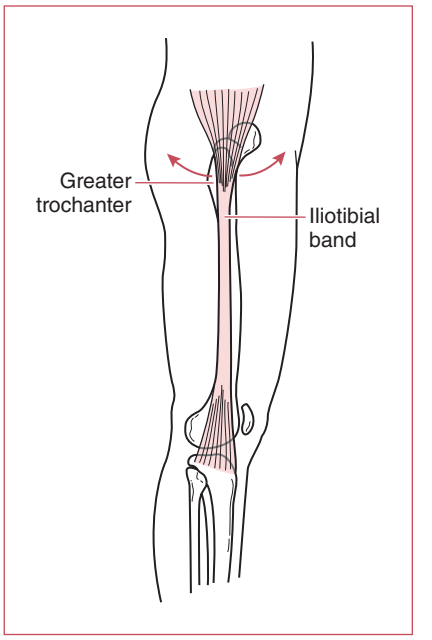

FIGURE 4–90 Iliotibial band (ITB) syndrome (lateral view).

HIP ADDUCTOR STRAIN (GROIN STRAIN)

General

- A common injury in sports, groin strain occurs due to resisted forceful abduction of the hip.
- The adductor groups are injured during eccentric contraction.
- Predisposing factors include relative weakness and tightness of the adductor muscle groups.
- It is important to distinguish muscle strain from adductor avulsion fracture.

Clinical Features

- Presents as pain in the adductors distal to their origin at the ramus or adductor tubercle.

PROVOCATIVE TEST

- Pain with resisted adduction and occasionally with hip flexion
- On palpation, there is tenderness of the adductor muscle.

Imaging

- Radiographs of the hip including the adductor tubercle to rule out avulsion

TREATMENT

- Rest, ice, NSAIDs, and then advance to stretching and strengthening

GREATER TROCHANTERIC HIP BURSITIS (FIGURE 4–91)

General

- Inflammation of the subgluteus maximus bursa, which is located over the greater trochanter and deep to the gluteus medius and gluteus minimus and TFL

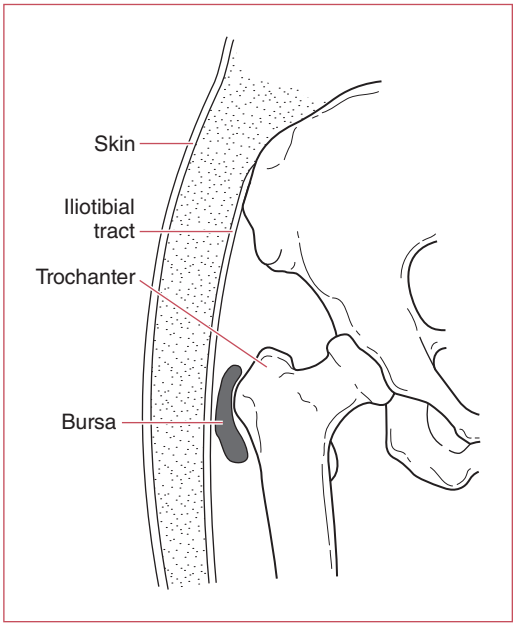

FIGURE 4–91 Greater trochanteric bursa. Note the relationship of the greater trochanteric bursa between the iliotibial band (ITB) and the greater trochanter of the hip (anterior view).

- It is associated with several conditions that cause altered gait mechanics, muscle imbalance, and reduced flexibility: Hip OA, obesity, leg length discrepancy, direct trauma, overuse, herniated lumbar disc, and hemiparesis.
- This condition may also cause external snapping hip syndrome.

Clinical Features

- Patients report pain at night and are unable to lie on the affected side.

PROVOCATIVE TEST

- Tenderness over the greater trochanter on palpation or during movement from full extension to flexion
- A snap may be palpable over the greater tubercle.

Imaging

- Radiographs of the hip to rule out bony pathology. In persistent cases, MRI may be indicated to rule out more significant pathology, such as a gluteus medius/minimus tear.

Treatment

- ITB stretching and NSAIDs. In severe cases, a cane may be needed for support and stability.
- Strengthening of the hip abductor muscles
- Local corticosteroid injection for resistant cases

🕮 SUPERIOR ILIAC CREST CONTUSION (HIP POINTER)

General

- Typically sustained from direct trauma to the superior iliac crest (as would occur in contact sports, for example); from direct trauma to the anterior aspect of the hip during a sporting event (especially American football, rugby, and hockey)
- The regional bone and soft tissue structures can be affected.
- Overall incidence is low.

Clinical Features

- Pain focal to the superior iliac crest
- Regional contusion/hematoma can occur.
- Weight bearing and hip ROM are generally painful.

Imaging

- Radiographs are reasonable for ruling out more significant injury to the bone (fractures/avulsions), especially in a younger patient population. Otherwise, radiographs are often without abnormality.
- US can be performed by a skilled sonographer to assess the iliac crest and extra-articular soft tissue structures.
- MRI may be appropriate in refractory cases to rule out alternative etiologies.

Treatment

- Conservative management is mainstay: Rest, ice, compression (controls hematoma formation/progression), acetaminophen.
- Local injection of anesthetic with or without steroid (however, data are limited)
- Return to play/normal activities within 2 to 4 weeks

POSTERIOR HIP DISLOCATION

General

- 🕮 This is the most common type of hip dislocation (90%).
- It may occur during an automobile accident when the hip is flexed, adducted, and medially rotated. The knee strikes the dashboard with the femur in this position, driving it posteriorly. In this position, the head of the femur is covered posteriorly by the capsule and not by bone.
- Due to the proximity of the sciatic nerve to the hip posteriorly, the sciatic nerve may be stretched or compressed in posterior hip dislocations.
- Note: Anterior hip dislocations may result in femoral nerve compromise.
- AVN may occur in 10% to 20% of patients.

Clinical Features

- The hip will be flexed, adducted, and internally rotated.
- The affected leg appears shorter because the dislocated femoral head is higher than the normal side.
- There will be an inability to abduct the affected hip.

Imaging

- Hip radiographs

Treatment

- This is an orthopedic emergency due to potential vascular compromise and sciatic nerve injury.

AVASCULAR NECROSIS OF THE FEMORAL HEAD (FIGURE 4–92)

General

- Also known as osteonecrosis of the hip or aseptic necrosis of the hip
- This condition is characterized by death of the femoral head without sepsis.
- Interruption of the vascular supply is the defining common pathway of the disease process.
- In children aged 2 to 12 years, this is known as Legg–Calvé–Perthes disease.
- The most common causes in adults are chronic corticosteroid use and alcohol abuse.

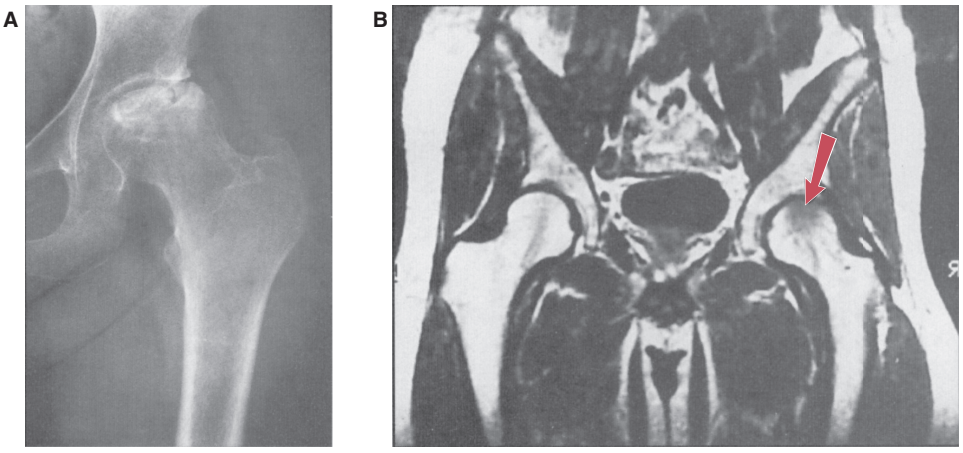

FIGURE 4–92 (A) X-ray of the left hip demonstrating sclerosis and flattening of the femoral head. (B) MRI scan reveals AVN of the left femoral head (arrow).
AVN, avascular necrosis.
Sources: From Cabanela ME. Hip arthroplasty in osteonecrosis of the femoral head. In: Urbaniak M, Jones JPM, eds. *Osteonecrosis: Etiology, Diagnosis, and Treatment*. American Academy of Orthopaedic Surgeons; 1997, with permission; Poss R, ed. *Orthopedic Knowledge Update 3*. American Academy of Orthopaedic Surgeons; 1990:540, with permission.

Clinical Features

- Pain may be present in the groin, anterior thigh, or even the knee.
- Pain is elicited on ROM and with weight bearing on the hip.
- Symptoms are of insidious onset.
- Shortened swing and stance phases on the affected side may be observed.
- There is loss of external and internal rotation of the hip. On hip flexion, the hip will externally rotate.

Imaging

- Irregular or mottled femoral head on plain films; femoral head collapse in later stages
- 🕮 MRI of both hips is indicated. MRI is most sensitive to early changes and is more specific than a bone scan:
  - There is low signal intensity on T1 imaging that may appear as rings, wedges, or irregular configurations.
  - T2 images may show a double line sign with a high signal intensity zone inside of a low signal intensity margin.

Treatment

- The main objective is to maintain the femoral head within the acetabulum while healing and remodeling occurs.
- Bracing and casting may help in the pediatric population to retain the femoral head within the acetabulum.
- Femoral head core decompression may be used to treat patients for earlier stages of AVN without compromise of femoral head morphology.
- Adults may require total hip arthroplasty (THA) in later stages of AVN with evidence of femoral head collapse.

🕮 HIP FRACTURES

General

- Osteoporosis of the hip carries an increased incidence of fracture.
- Osteoporosis of the hip is associated with both nonmodifiable and modifiable risk factors:
  - Nonmodifiable risk factors include age, sex, and race:
    - Approximately 60% of hip fractures occur in patients >75 years of age.
    - Females have higher incidence of hip fracture than males.
    - Among females, there is a 2 to 3:1 higher rate of fracture in European Americans than in African Americans.
  - Modifiable risk factors:
    - Alcohol and caffeine consumption
    - Smoking
    - Medications (steroids, antipsychotics, benzodiazepines)
    - Malnutrition
    - Body weight <90% of ideal
- Surgery for hip fracture and degeneration carries considerable risks of morbidity and mortality:
  - Venous thrombosis occurs in >50% of unprotected patients. Note: The risk for pulmonary embolism is highest during the second and third week.
  - The incidence of heterotopic ossification is high (>50%) after total hip replacement and is the most common complication, although <10% lose ROM.
  - The mortality rate for those who survive a hip fracture is 20% to 30% after 1 year and approximately 40% after 2 years.
  - Approximately 50% of patients return to their premorbid level of functioning.
- Classification is based on the anatomy of the proximal femur and consists of three main types: Intracapsular, intertrochanteric, and subtrochanteric **(Figure 4–93)**. See the following sections for further details.

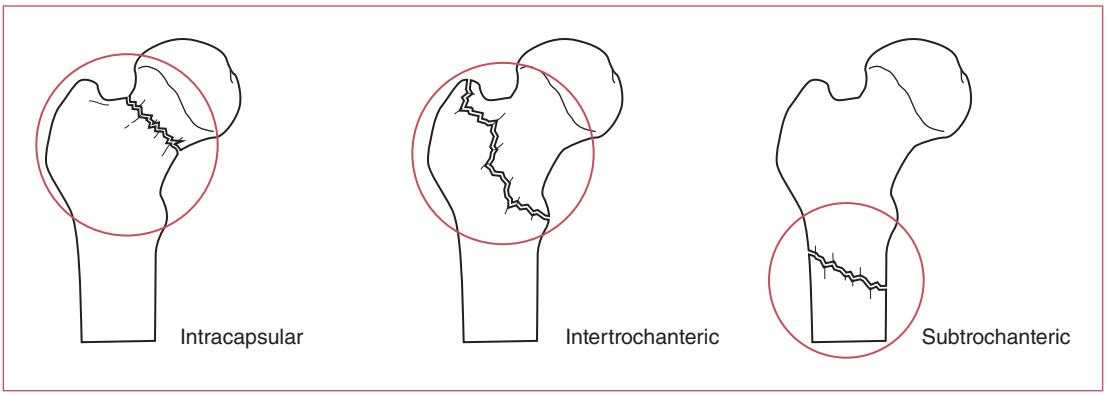

FIGURE 4–93 Classification of hip fractures based on the anatomy of the proximal femur. Intracapsular is synonymous with the femoral neck/subcapital fracture.

INTRACAPSULAR/FEMORAL NECK FRACTURES

General

- Fractures of the femoral neck are classified by Garden classification **(Figure 4–94)**:
  - Stage I: Incomplete, nondisplaced with occasional valgus angulation
  - Stage II: Complete, nondisplaced, occasionally unstable
  - Stage III: Displaced with the hip joint capsule partially intact
  - Stage IV: Displaced with the hip joint capsule completely disrupted
- Morbidity associated with the fracture involves disruption of the blood vessels to the femoral head causing osteonecrosis.
- Postoperatively, complications can include nonunion or osteonecrosis.

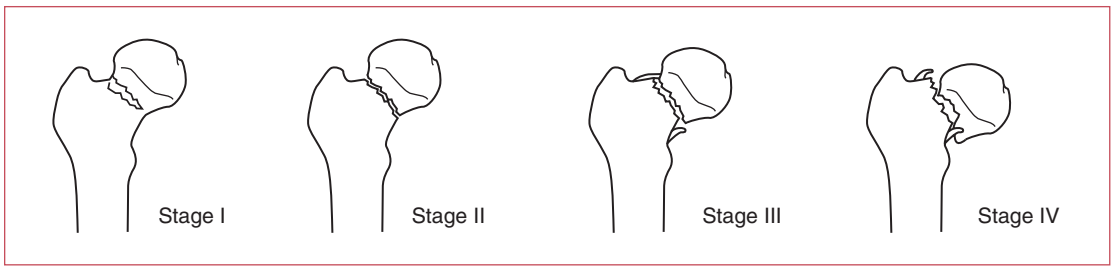

FIGURE 4–94 Garden classification of femoral neck fractures.

Clinical Features

- Common presentation of hip fracture is a patient with hip pain, limb externally rotated greater than normal, and an apparent shortened limb on the affected side.

Imaging

- Radiographs of the hip are essential.
- Bone scan may be necessary for occult fractures.
- MRI can be an additional imaging option.

Treatment

- Garden Stages I and II:
  - *Surgical:* Pins across the fracture site or a cannulated hip screw is used for stabilization.
  - *Conservative:*
    - May be treated conservatively if the patient is unfit for surgery or in the case of an old, impacted fracture
    - Rehabilitation early with partial or full weight bearing

- Garden Stages III and IV:
  - *Surgical intervention:*
    - Replacement of the femoral head using cemented or noncemented hemiarthroplasty, total hip replacement, or bipolar arthroplasty
    - Bipolar implants consist of a femoral component that articulates by snap-fit into a cup that moves freely within the acetabulum.
    - Theoretically reduces friction and impact at the prosthetic–acetabular junction by allowing motion at two sites: Prosthetic inner-bearing surface and prosthetic-bearing acetabular surface
    - Acetabular erosion and protrusion should theoretically be reduced by reducing motion at the acetabulum.
  - *Postop rehabilitation:*
    - Rehabilitation of cemented cases by immediate full weight bearing
    - Rehabilitation of uncemented cases may be partial or full weight bearing.
    - There are posterior and anterior approaches to THA. The posterior approach is more at risk for dislocation.
    - 🕮 In a posterior THA approach, total hip precautions must be followed to avoid dislocation. Avoid hip flexion over 90 degrees, hip adduction past midline, and extreme hip internal rotation:
    - A high chair height is preferred to a low height in order to reduce hip flexion and potential for posterior hip dislocation.
    - 🕮 After an anterior approach THA, different precautions are required, including avoidance of hip extension and external rotation (i.e., opposite of posterior THA precautions).
    - Dislocation after hip replacement occurs at varying frequency. Prompt reduction under general anesthesia will most likely preserve the replacement.
- Sepsis is a cause of dislocation after hip replacement and must be considered.

INTERTROCHANTERIC HIP FRACTURES (FIGURE 4–95)

🕮 General

- This is the most common type of hip fracture.
- Highly fragmented fractures may result in significant blood loss and hypovolemia.
- Postoperatively, a leg length discrepancy may result due to comminution.
- Moderately high forces are generated in this area, and a strong fixation is required.
- Fractures may be nondisplaced, displaced two-part fractures, or unstable three-part fractures.

Clinical Features

- The common presentation of hip fracture is hip pain with an externally rotated and shortened limb.

Imaging

- Radiographs and possible CT scan or MRI

Treatment

- *Surgical:*
  - A compression screw or angle nail plate may be used.
  - If the fixation is unstable, medial displacement osteotomy of the femur may be required.
- *Rehabilitation:*
  - Progress from partial to full weight bearing

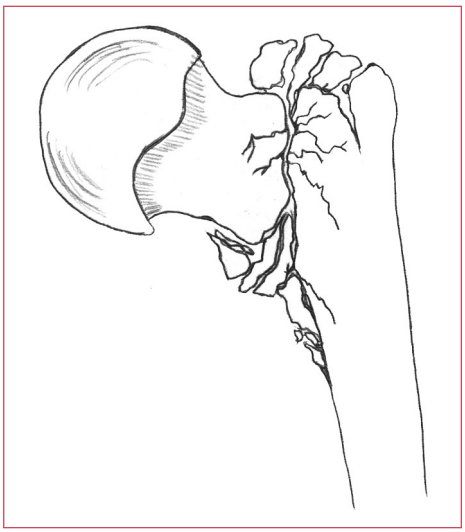

FIGURE 4–95 Intertrochanteric hip fracture. (Note the bone fragmentation.)

SUBTROCHANTERIC HIP FRACTURES (FIGURE 4–96)

General

- This region is subjected to very high mechanical stresses and is the most difficult to stabilize surgically.
- Fractures may be simple, fragmented, or comminuted.

Clinical Features

- The typical signs and symptoms of hip fracture are present: Hip pain with an external rotated limb with possible shortening and malalignment.

Imaging

- Radiographs of the hip and possible CT scan

Treatment

- *Conservative:*
  - In these fractures, rehabilitation may be delayed until fracture healing is evident.
  - Weight bearing progresses from partial to full.
- *Surgical:*
  - ORIF will be necessary with several choices of fixation hardware.
  - A blade plate and screws may be sufficient in some cases.
  - An intramedullary (IM) rod may be required to make an extremely strong fixation through the proximal femur and trochanteric region.

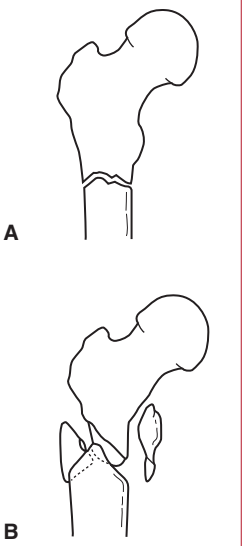

FIGURE 4–96 Subtrochanteric femur fractures. (A) Simple. (B) Comminuted.

FEMORAL NECK STRESS FRACTURES

General

- There are two basic types of femoral neck fractures: Compression and transverse.
- **Compression-type fractures** are more common and generally occur along the inferior neck of the femur. They are more stable.
- **Transverse-type fractures** occur with the fracture along the superior aspect of the femur neck. These are more unstable and are also termed *tension side fractures*.
- Endurance athletes, such as runners, triathletes, and military recruits, are susceptible to proximal femoral stress fractures.

Clinical Features

- Femoral neck stress fractures present as groin pain that is made worse with ADLs.
- There will often be pain at extreme ranges of internal and external rotation.

Imaging

- Radiographs may be negative at first and in time may show periosteal thickening or a radiolucent line.
- If plain films are negative, a bone scan should be done.
- Bone scans may be positive 2 to 8 days after onset of symptoms.

Treatment

- Compression type:
  - Because they are more stable, they may be treated with bedrest.
  - When there is no pain at rest, then weight bearing to limitation of pain is allowed.
  - If compression-type fractures progress, they may require internal fixation.
- Transverse or tension type:
  - These fractures are generally treated with ORIF due to the high risk of displacement.

SLIPPED CAPITAL FEMORAL EPIPHYSIS (FIGURE 4–97)

General

- This is an injury to the epiphyseal growth plate at the head of the femur causing displacement of the plate.
- The common ages of incidence are children aged 11 to 16 years.
- The injury may be associated with a direct hip trauma.

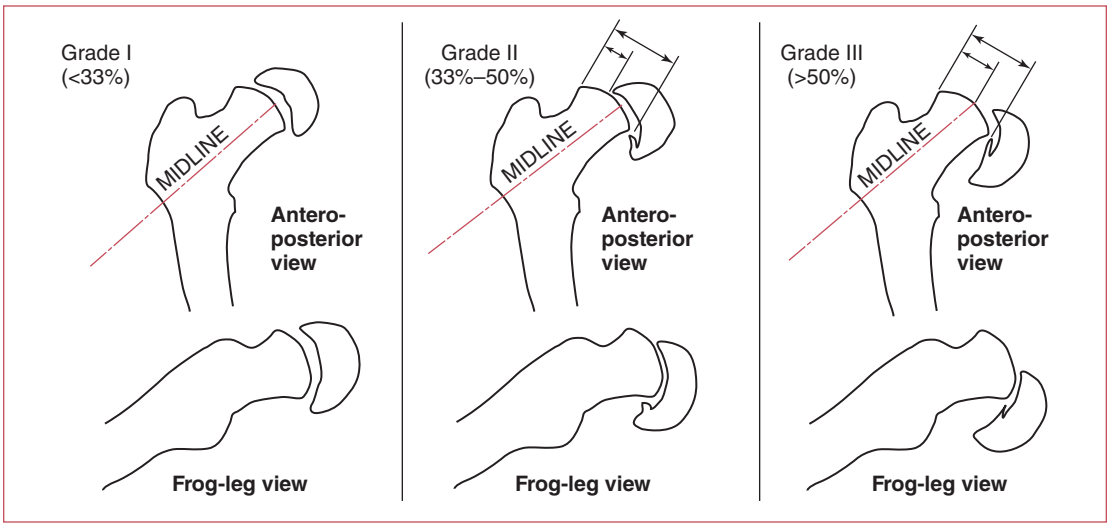

FIGURE 4–97 SCFE classification: Grade I, Grade II, and Grade III (% indicates degree of slippage for each grade). SCFE, slipped capital femoral epiphysis.

Clinical Features

- SCFE typically presents with groin or hip pain but may also present as thigh or knee pain.
- It is often associated with an antalgic gait.
- Internal hip rotation is limited, and the extremity externally rotates when the hip is flexed.
- Muscle spasms and synovitis occur in the acute phase.

Imaging

- Hip radiographs and/or CT will reveal medial and posterior displacement of the epiphysis.

Treatment

- Immediate non-weight-bearing status
- Surgical stabilization is required.
- Endocrine testing should be done to rule out:
  - Growth hormone deficiency
  - Hyperthyroidism
  - Hypothyroidism
  - Panhypopituitarism
  - Multiple endocrine neoplasia syndromes

AVULSION FRACTURES

Ischial Tuberosity Avulsion Fractures

General

- Ischial tuberosity avulsion fractures generally occur with a forceful hamstring contracture with the knee in extension and the hip in flexion.
- The origin of the hamstring is pulled away from the ischial tuberosity.

Clinical Features

- Ischial tuberosity avulsion fractures present with acute-onset pain and tenderness over the ischial tuberosity.
- The patient may describe a "popping" or tearing sensation with a palpable defect on examination.
- Ischial avulsion must be differentiated from ischial bursitis:
  - Ischial tuberosity bursitis (also known as weaver's bottom) is of insidious onset and is more progressive in presentation.
- There will be reproduction or increased pain on straight leg raise (hamstring stretching).

Imaging

- Radiographs should reveal the avulsion at the ischium.

Treatment

- Generally, this avulsion is treated with rest, ice, and weight bearing as tolerated.
- After achieving full ROM, resistance exercises can be started.
- Surgery may be required for a displaced apophysis.
- Rarely, there can be significant scar formation around the sciatic nerve in the posterior thigh.

Anterior Superior Iliac Spine Avulsion Fractures

General

- The ASIS is the point of attachment for the sartorius muscle.
- This avulsion is caused by forceful contraction (e.g., kicking, running, jumping) with the hip extended and the knee flexed.
- If the lateral femoral cutaneous nerve is involved, paresthesias may involve the anterolateral thigh.

Clinical Features

- Tenderness is present over the ASIS.
- There should be pain on hip flexion.

Imaging

- Radiographs should demonstrate the avulsion fracture.

Treatment

- ASIS avulsions are generally treated with rest, ice, and weight bearing as tolerated.
- They may require the knee to be splinted in flexion to reduce tension on the avulsion segment.
- Stretching and strengthening are instituted with reduction in pain.
- Surgery may be required for a displaced apophysis.

Anterior Inferior Iliac Spine Avulsion Fractures

General

- The AIIS is the point of origin for the rectus femoris muscle.
- Avulsions here may also be caused by forceful kicking and contraction of the quadriceps.

Clinical Features

- AIIS avulsion fractures can present with acute-onset pain over the AIIS or groin pain.
- Pain will be produced with quadriceps contraction, hip flexion, or hip extension.

Imaging

- Radiographs should demonstrate the avulsion.

Treatment

- These avulsions are generally treated with rest and ice with weight bearing as tolerated.
- Strengthening may begin after full ROM is achieved.
- Surgery may be required for a displaced apophysis.

OSTEITIS PUBIS

General

- This is an inflammatory condition of the joint of the pubic rami.
- It is often caused by cumulative overuse of the adductors.

Clinical Features

- Osteitis pubis presents as pubic symphysis or groin pain and may radiate into the thigh.
- Normal ambulation may produce a popping in the pubic region.
- Pain will be produced on resisted adduction.
- Pain can be elicited in the groin or pubic area with one-legged hopping.

Imaging

- Radiographs and/or CT may show periosteal thickening.

Treatment

- Generally, rest and NSAIDs are the first-line treatments.
- Corticosteroid injection may be required in persistent cases.
- Stretching and strengthening may proceed after reduction in pain.
- Surgery for arthrodesis may be required in severe cases.

MYOSITIS OSSIFICANS

General

- Myositis ossificans is the formation of heterotopic ossification within muscle.
- Ossification within an area of muscle forms from encapsulated blood secondary to a hematoma.
- It is usually the result of repeated trauma to that area of muscle or can be due to a direct blow to the hip.
- The quadriceps is the most common location.
- Other sites include the brachialis, deltoid, intercostal space, erector spinae, pectoralis, and gluteal muscles
- US, heat, massage, or repeated trauma at the onset of myositis ossificans can exacerbate the process.

Clinical Features

- There will be pain and a palpable mass at the site of the lesion.
- If the ossifying mass involves a nerve, related nerve impingement symptoms may occur.

Imaging

- Initially radiographs will reveal a soft tissue mass.
- Calcific flocculations can develop within 14 days.
- Ossification can be seen between 2 and 3 weeks.
- Bone scan and MRI are more sensitive than radiographs in the early stages. US can be helpful for office or bedside diagnosis of a hematoma and myositis ossificans.

Treatment

- Gentle ROM
- Prevention of contractures is of prime importance.

- Strengthening of the involved muscles should be done progressively.
- Surgery may be necessary in cases resulting in nerve entrapment, decreased ROM, or loss of function.
- If possible, surgery should be delayed until the lesion matures at 10 to 12 months.
- Radiation therapy can be tried for recalcitrant symptoms.

LOWER EXTREMITIES: THE KNEE

KNEE FUNCTIONAL ANATOMY

- The knee joint is a modified hinge joint.
- The knee is the largest joint in the body. It is susceptible to injury because it is at the end of the tibia and femur, two long lever arms.

KNEE RANGE OF MOTION

ADULT KNEE ROM	DEGREES
Knee flexion	135
Knee extension	0
Knee internal rotation	10
Knee external rotation	10

ROM, range of motion.

MUSCLES (FIGURES 4–98 AND 4–99)

Knee Extensors

- Quadriceps (femoral nerve: L2, L3, L4):
  - Rectus femoris
  - Vastus lateralis
  - Vastus intermedius
  - Vastus medialis obliquus
- All join with the quadriceps tendon.
- The vastus medialis and lateralis draw the patella in their respective directions.

Knee Flexors

KNEE FLEXORS	SPINAL INNERVATION	PERIPHERAL INNERVATION
Semitendinosus	L5, S1, S2	Sciatic nerve (tibial division)
Semimembranosus	L5, S1, S2	Sciatic nerve (tibial division)
Biceps femoris, long head	L5, S1, S2	Sciatic nerve (tibial division)
Biceps femoris, short head	L5, S1, S2	Sciatic nerve (common peroneal/fibular division)
Gracilis	L2, L3, L4	Obturator nerve
Gastrocnemius	S1, S2	Tibial nerve
Sartorius	L2, L3	Femoral nerve

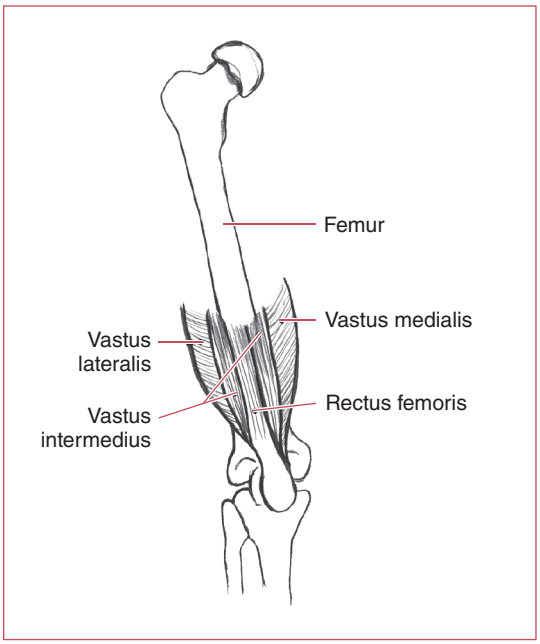

FIGURE 4–98 Anterior and medial aspects of the thigh (deeper muscles).

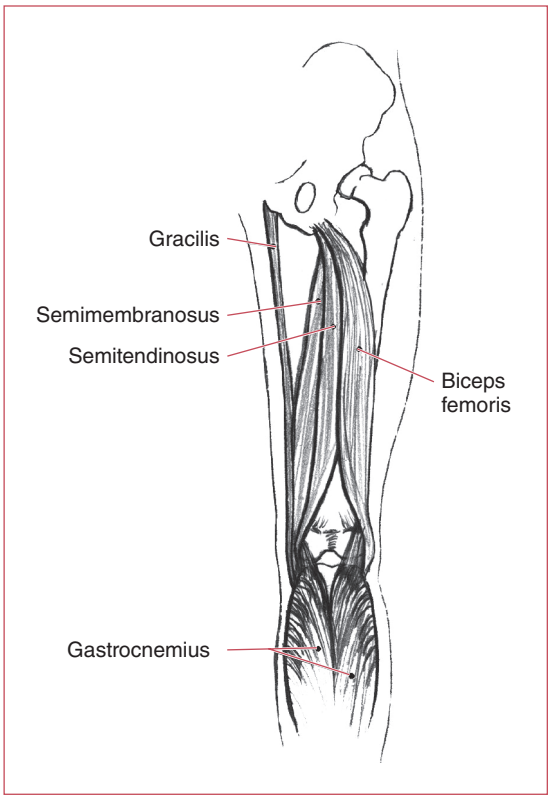

FIGURE 4–99 Posterior muscles of the thigh. (Sartorius is not shown.)

Medial Rotators

KNEE MEDIAL ROTATORS	SPINAL INNERVATION	PERIPHERAL INNERVATION
Semitendinosus	L5, S1, S2	Sciatic nerve (tibial division)
Semimembranosus	L5, S1, S2	Sciatic nerve (tibial division)
Sartorius	L2, L3	Femoral nerve
Gracilis	L2, L3, L4	Obturator nerve

Lateral Rotators (Figure 4–99)

KNEE LATERAL ROTATORS	SPINAL INNERVATION	PERIPHERAL INNERVATION
Biceps femoris, long head	L5, S1, S2	Sciatic nerve (tibial division)
Biceps femoris, short head	L5, S1, S2	Sciatic nerve (common peroneal/fibular division)

Other

- Popliteus (tibial nerve: L4, L5, S1)
  - Unlocks the knee

Bony Anatomy

- The knee can be divided into the medial femoral–tibial, lateral femoral–tibial, and patellofemoral compartments.

Femoral–tibial joint:

- The primary motion is flexion and extension.
- The secondary motion is axial rotation when the knee is flexed.
- The basic motion of the femur is rolling and gliding.

Tibial plateau:

- Characterized by curved surfaces corresponding to the medial and lateral femoral condyles
- A raised prominence between the curved surfaces helps prevent rotation in extension.
- The lateral tibial plateau is convex in shape. This allows the lateral femoral condyle to move farther backward than the medial condyle. This causes internal tibial rotation with flexion.

Patella:

- The patella is a sesamoid bone embedded in the extensor muscle/tendon unit.
- Efficiency of the knee extensor muscles is increased by 150% due to the mechanical advantage provided by the patella.
- The articular surface of the patella has two facets divided by a ridge, which helps with tracking over the condyles.

- 🕮 The Q angle **(Figure 4–100)** is formed by the long axes of the femur and the tibia and reflects the natural valgus attitude of the knee:
  - Males approximately 13 degrees
  - Females approximately 18 degrees
- Excessive valgum of the knee is termed *genu valgum* (knock-kneed).
- Excessive varum of the knee is termed *genu varum* (bow-legged).
- Hyperextension at the knees is termed *genu recurvatum* (back-kneed).

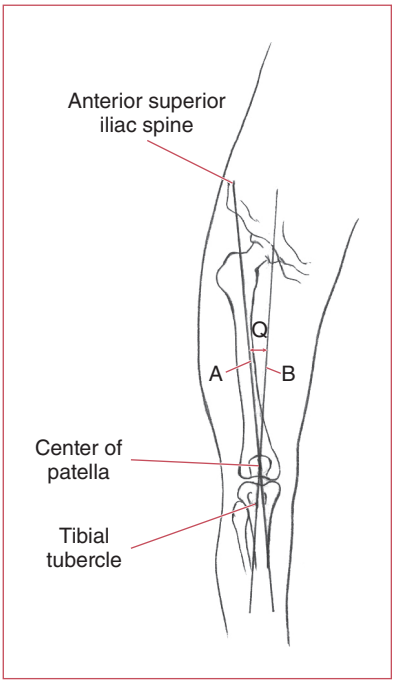

FIGURE 4–100 Q angle.

LIGAMENTS OF THE KNEE (FIGURES 4–101 TO 4–103)

Anterior Cruciate Ligament

FUNCTIONAL ANATOMY

- The anterior cruciate ligament (ACL) originates at the posteromedial aspect of the lateral femoral condyle, travels anteromedially through the intercondylar notch, and attaches to the medial tibial eminence:
  - The ACL runs inferior, anterior, and medial in direction to its tibial attachment. (Imagine putting your hands in your front pockets. That diagonal direction coincides with the pathway of the ACL.)
- 🕮 Its primary function is to limit anterior tibial translation.

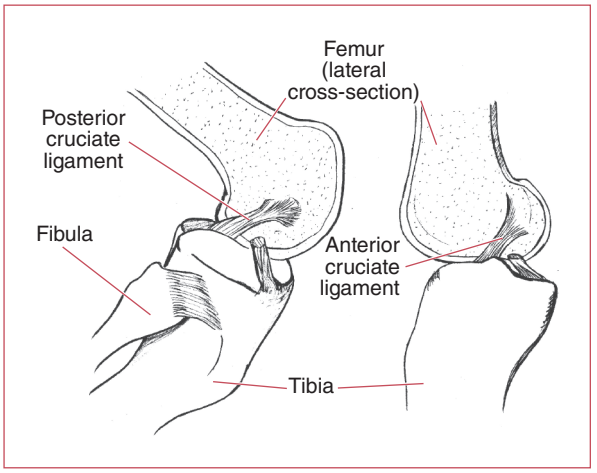

FIGURE 4–101 Anterior and posterior cruciate ligaments (ACL, PCL) of the knee.

- It also prevents posterior translation of the femur and hyperextension of the knee.
- In addition, it limits internal rotation of the femur when the foot is fixed and the knee is locked.
- The ACL tightens with full extension and loosens in flexion.
- Femoral external rotation loosens the ACL, and internal rotation tightens it.
- 🕮 In flexion, it draws the femoral condyles anteriorly.
- ACL-deficient knees create increased pressures on the posterior menisci.

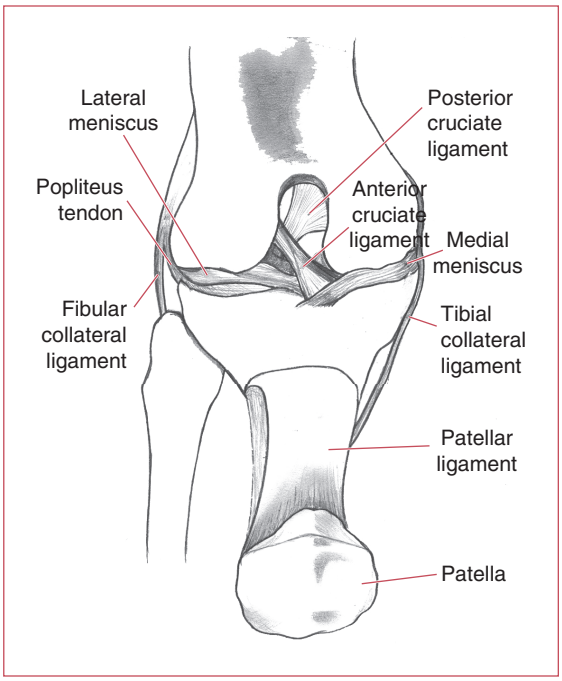

FIGURE 4–102 Anterior view—ligaments of the knee.

Posterior Cruciate Ligament

FUNCTIONAL ANATOMY

- The posterior cruciate ligament (PCL) originates from the anterolateral aspect of the medial femoral condyle, through the intercondylar notch, and inserts onto the posterior aspect of the tibial plateau.
- The PCL runs inferior, posterior, and lateral in direction to its tibial attachment. (Imagine putting your hands in your back pockets, or more simply, opposite of the fiber directions of the ACL.)
- Its primary function is to restrain posterior tibial translation.
- The ligament is looser in extension and tighter in flexion.
- In extension, the PCL pulls the femur posteriorly.
- PCL-deficient knees place more force on the patellofemoral joint.

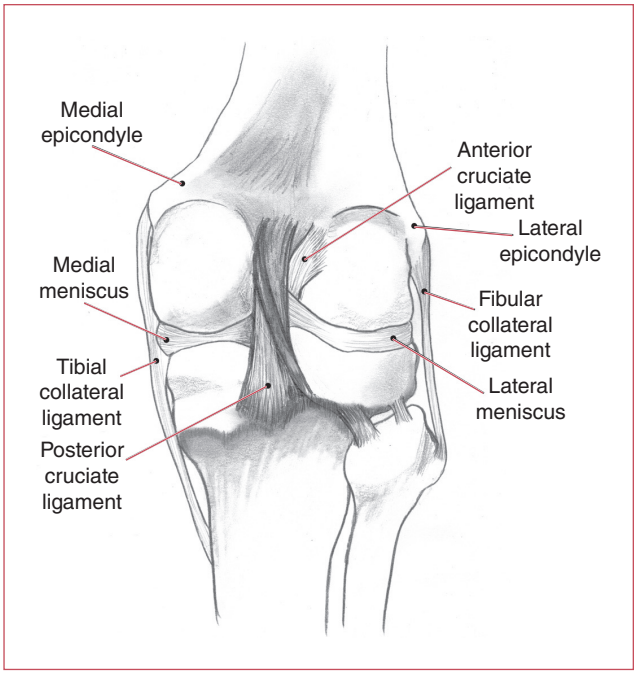

FIGURE 4–103 Posterior view—ligaments of the knee.

Medial Collateral Ligament

FUNCTIONAL ANATOMY

- The medial collateral ligament (MCL) originates at the medial femoral condyle and inserts onto the superior aspect of the medial tibia.
- 🕮 It has an attachment to the medial meniscus.
- In full extension, the MCL tightens to full tension.
- Tension is increased with abduction stress at increasing positions of flexion.
- It restrains valgus stresses on the knee.

Lateral Collateral Ligament

FUNCTIONAL ANATOMY

- The lateral collateral ligament (LCL) originates from the lateral femoral condyle posteriorly and superiorly and attaches to the superior part of the lateral fibula.

- It does not have an attachment to the lateral meniscus.
- It restrains varus stresses on the knee.
- Peak stress is achieved with adduction when the knee is at 70-degree flexion.

Capsular Ligaments (Figure 4–104)

- In general, the posterior capsule resists knee hyperextension.

OBLIQUE POPLITEAL LIGAMENT

- The oblique popliteal ligament (OPL) arises from the semimembranosus tendon.
- It strengthens the fibrous posterior capsule of the knee joint.
- It resists knee extension.
- The OPL is attached to the capsule and lateral meniscus.

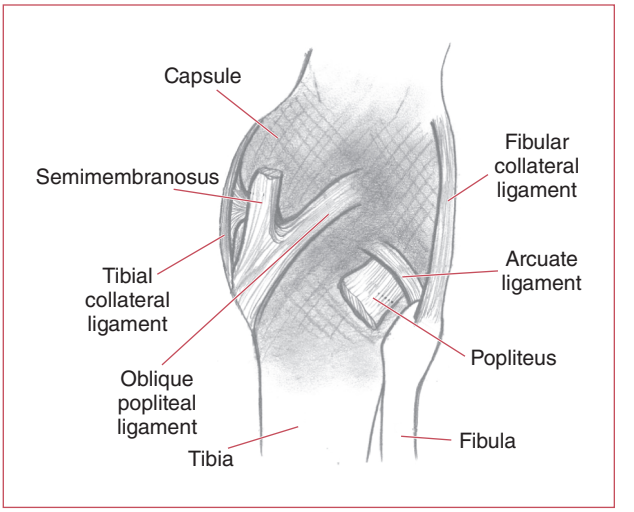

FIGURE 4–104 Knee joint—capsular ligaments.

Arcuate Popliteal Ligament Complex

- The arcuate popliteal ligament complex (APLC) provides attachment for the posterior horn of the lateral meniscus.
- It reinforces the lateral aspect of the knee and gives posterolateral rotatory stability.
- It also provides restraint to posterior tibial translation.
- Its attachment can be mistaken for a tear of the posterior horn of the lateral meniscus on MRI.

MENISCI OF THE KNEE (FIGURE 4–105)

- The two menisci (medial and lateral) are composed of crescent-shaped fibrocartilaginous tissue.
- They deepen the articular surface area of the tibia to provide more stability for the femoral condyles and increased force dispersion to the tibial plateau.

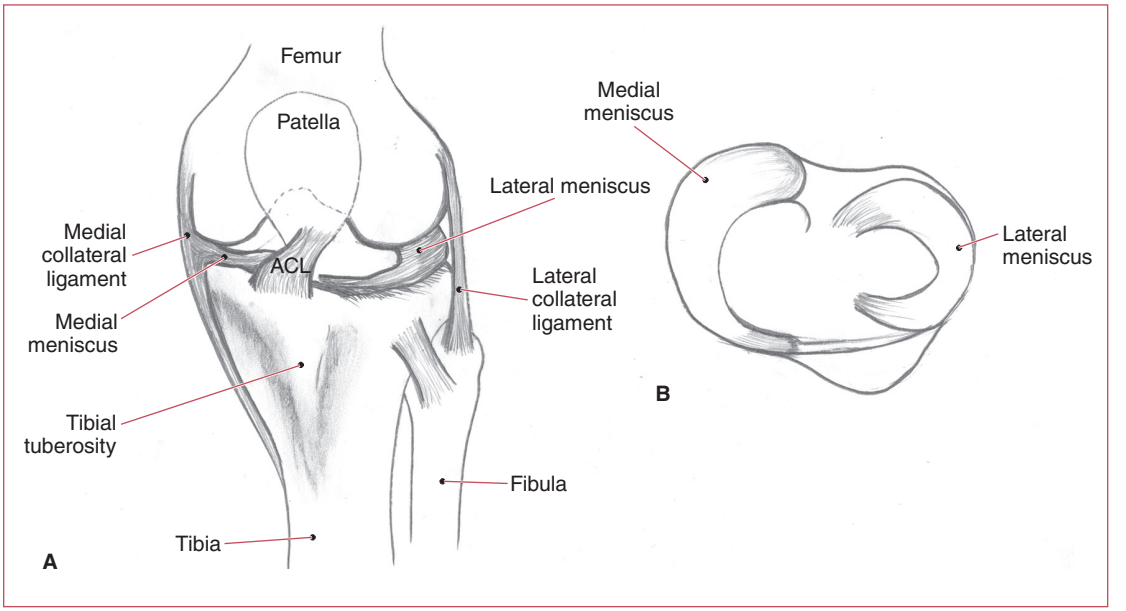

FIGURE 4–105 Major structures of the knee joint. (A) Anterior view. (B) View from above with the femur removed.

- The peripheral outer one-third of the menisci is well vascularized.
- The inner two-thirds are poorly vascularized and usually cannot be surgically repaired.

Medial Meniscus

- The medial meniscus is larger and longer than the lateral meniscus **(Figure 4–105B)**.
- It is "C"-shaped.
- The peripheral border is attached to a portion of the MCL. Medial knee injuries can result in injuries to both the MCL and the medial meniscus.

Lateral Meniscus

- The lateral meniscus is "O"-shaped.
- It covers a larger area than the medial meniscus.
- It is joined to the medial femoral condyle by the posterior meniscofemoral ligament.

BURSAE OF THE KNEE (FIGURE 4–106)

Anterior

- Prepatellar bursae:
  - Located between the skin and the anterior patella
  - It is the most commonly damaged bursa.
  - Inflammation of this bursa is caused by prolonged kneeling. Also known as housemaid's knee.
- Suprapatellar bursa:
  - This bursa is located between the quadriceps femoris muscle and the femur and usually communicates with the joint capsule.

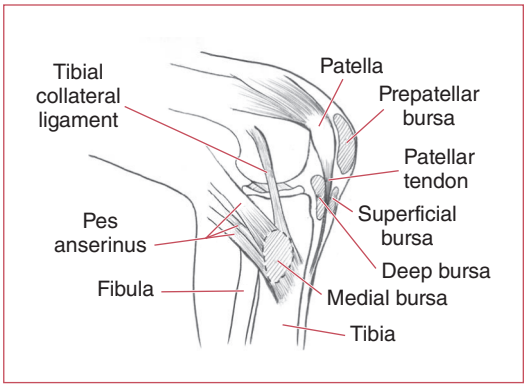

FIGURE 4–106 Bursae of the knee that are clinically significant (medial view of knee).

- Deep infrapatellar bursa is located between the patellar tendon and the tibia.
- Superficial or subcutaneous infrapatellar bursa:
  - This bursa is located between the skin and the tibial tuberosity.
  - Associated with kneeling in an upright position (e.g., kneeling on a prayer bench). Inflammation of the bursa is also known as vicar's knee.

Lateral Bursae

- There is a lateral bursa located between the biceps muscle and fibular collateral ligament.
- A second lateral bursa is located between the popliteus muscle and the fibular collateral ligament.
- A third lateral bursa is located between the lateral femoral condyle and popliteus muscle.

Medial Bursae

- Pes anserinus bursa:
  - Located between the tendons of the **S**artorius, **G**racilis, and semi**T**endinosus muscles ("**S**ay **G**race before **T**ea") and MCL.
  - Pes anserine bursitis may be misdiagnosed as a medial meniscal lesion when swollen and painful.
- A second medial bursa is located between the MCL and the semitendinosus.
- A third medial bursa is located between the semimembranosus and the tibia.

Posterior Bursae

- There is a posterior bursa located between the medial head of the gastrocnemius and capsule and extending under the semimembranosus:
  - It communicates with the joint cavity.
  - It can be irritated by abnormal strain.

  - When distended, it is known as a *Baker's cyst*.
  - A Baker's cyst is a distension of the bursa and outpocketing of the synovial membrane in this location. It can mimic a deep vein thrombosis due to the increased warmth, leg girth, and associated pain.
- There is also a posterior bursa located between the lateral head of the gastrocnemius and the capsule.

CLINICAL TESTS FOR THE KNEE (Malanga & Nadler, 2005)

Tests for Meniscal Injuries

Joint Line Palpation

- While this is a basic clinical test, palpation of the medial and lateral joint lines can help detect anterior horn meniscal injuries.
- Greater sensitivity than McMurray's test
- Caution—joint line tenderness is commonly seen in other clinical diagnoses as well.

McMurray's Test (Figure 4–107)

- This test is used to diagnose meniscal tears and is especially useful for diagnosing posterior meniscal tears:
  - The patient lies supine with the knee flexed **(Figure 4–107A)**.
- The examiner palpates the medial joint line with their fingers and the lateral joint line with their thumb.
- With the knee flexed, rotate the tibia internally and externally on the femur **(Figure 4–107B)**.

TO TEST FOR MEDIAL MENISCAL DAMAGE

- With the knee flexed and the tibia externally rotated, push on the lateral side while applying valgus stress to the medial side of the knee joint **(Figure 4–107C).**
- Externally rotating the tibia while extending the knee stresses the medial compartment for testing the medial meniscus.
- With the leg externally rotated and in valgus, slowly extend the knee.
- A positive test for a torn medial meniscus (usually in the posterior region) yields an audible or palpable click and pain at the medial joint line **(Figure 4–107D).**

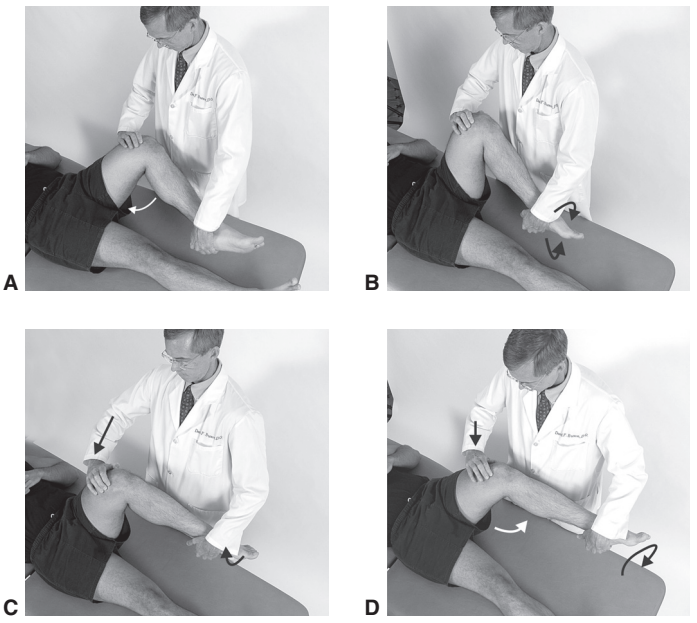

FIGURE 4–107 (A) McMurray's test: Patient lies supine with the knee flexed. (B) With a flexed knee, internally and externally rotate the tibia on the femur. (C) Place a valgus stress on the knee with the leg externally rotated. (D) Slowly extend the leg with the leg externally rotated and in the valgus position. If a click is heard or palpated, the test is positive for a torn medial meniscus, usually in the posterior position.
Source: Courtesy of JFK Johnson Rehabilitation Institute, 2000.

TO TEST FOR LATERAL MENISCAL INJURY

- Apply a varus stress and internally rotate the tibia while extending the knee.
- Internally rotating the tibia stresses the lateral compartment for testing the lateral meniscus.
- A positive test for a torn lateral meniscus (usually in the posterior horn) yields an audible or palpable click and pain.

Apley's Grind Test (Figure 4–108)

- This test is also used to diagnose a meniscus tear.
- The patient lies prone with the knee flexed to 90 degrees.
- The examiner places force downward on the heel, compressing the menisci between the femur and tibia.
- Pressure is maintained downward as the tibia is rotated.
- A positive test should elicit pain and indicates the injury is more likely meniscal than ligamentous; ask the patient to localize the pain to the medial or lateral compartment.

FIGURE 4–108 Apley's grind test.
Source: Courtesy of JFK Johnson Rehabilitation Institute, 2000.

Apley's Distraction Test (Figure 4–109)

- This is a test for collateral ligamentous (MCL or LCL) damage.
- The patient lies prone with the knee flexed.
- Traction force is applied while rotating the tibia internally and externally.
- Pressure on the menisci is reduced during this maneuver.
- A positive test elicits pain and indicates that the injury is more likely ligamentous rather than meniscal.

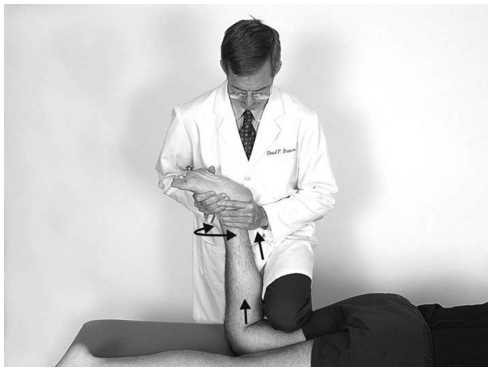

FIGURE 4–109 Apley's distraction test.
Source: Courtesy of JFK Johnson Rehabilitation Institute, 2000.

Bounce Home Test (Figure 4–110)

- This is a test for decreased knee extension.
- With the patient supine, the examiner flexes the knee while holding the heel **(Figure 4–110A)**.
- The knee is allowed to extend passively and should have a definitive endpoint. It should "bounce home" into extension **(Figure 4–110B)**.

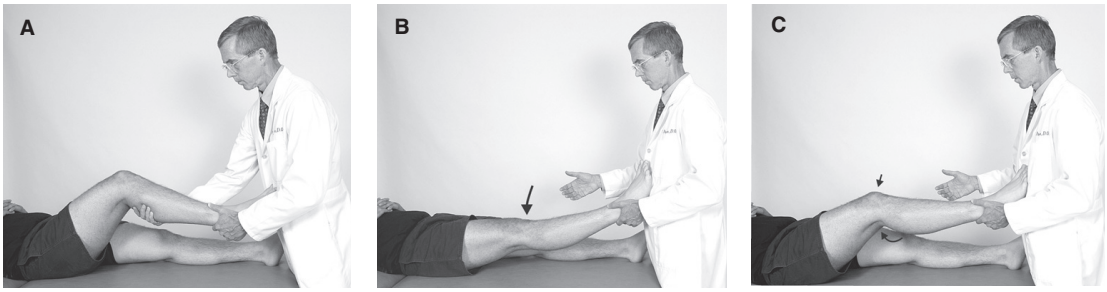

FIGURE 4–110 Bounce home test. (A) Examiner flexes the knee. (B) Examiner lets the knee passively extend. (C) Positive test occurs when full extension cannot be attained.
Source: Courtesy of JFK Johnson Rehabilitation Institute, 2000.

- A positive test occurs when full extension cannot be attained, and rubbery resistance is felt (**Figure 4–110C**).
- Causes for a positive test include a torn meniscus, loose body, intracapsular swelling, or fluid in the knee joint.

Test for Patellofemoral Disorders

Patellofemoral Grind Test (Figure 4–111)

- This test evaluates the quality of the patella articulating surfaces.
- The patient is supine with the legs in a neutral position.
- The examiner pushes the patella distally and then asks the patient to contract the quadriceps against the resistance on the patella. The patella should glide smoothly in a cephalad direction.
- A positive test will yield pain and crepitation on patella movement.

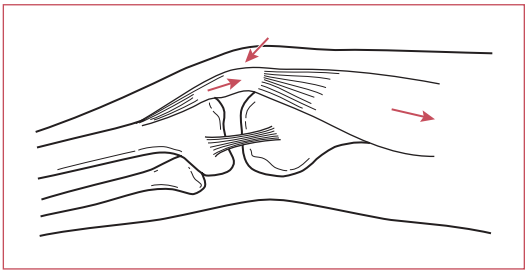

FIGURE 4–111 Patellofemoral grind test.

Tests for the Anterior Cruciate Ligament (ACL)

Lachman's Test (Figure 4–112)

- This test is used to assess ACL integrity and anterior knee integrity.
- Anterior tibial translation is evaluated.
- The patient is supine. The knee is flexed at 15 to 30 degrees. The examiner grasps the distal femur with one hand and the proximal tibia with the other hand.
- The femur is stabilized, and anterior force is applied to the tibia.
- 🕮 A positive test yields significant anterior movement with no distinct endpoint.
- A partial ACL tear may yield a test with a soft endpoint.

FIGURE 4–112 Lachman's test.
Source: Courtesy of JFK Johnson Rehabilitation Institute, 2000.

Anterior Drawer Test (Figure 4–113)

- The anterior drawer primarily tests the integrity of the ACL.
- With the patient supine, the knees flexed to 90 degrees, and the foot stabilized by the examiner's body, the examiner grasps the patient's knee with the fingers in the area of the hamstring insertions and the thumbs on the medial and lateral joint lines. The tibia is drawn toward the examiner (**Figure 4–113**).
- A positive test occurs when the tibia slides from under the femur with no distinct endpoint. Some anterior translation is normal. A distinct endpoint should be encountered in a normal exam (**see Figure 4–112**).

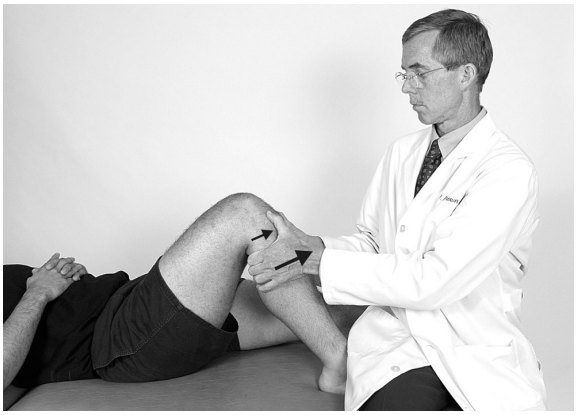

FIGURE 4–113 Anterior drawer test. The tibia is drawn toward the examiner. Positive anterior draw sign: Torn anterior cruciate ligament.
Source: Courtesy of JFK Johnson Rehabilitation Institute, 2000.

- External rotation tightens the posteromedial portion of the joint capsule, thereby restricting anterior movement. If the anterior translation with the leg in the neutral position is equal to the anterior movement with the leg externally rotated, the posteromedial joint capsule and the ACL may be damaged.
- Internal rotation tightens the posterolateral joint capsule. If the anterior translation with the leg in the neutral position is equal to the anterior movement with the leg internally rotated, the posterolateral joint capsule and the ACL may be damaged.
- 🕮 The test is not very sensitive because hemarthrosis, hamstring spasm, and other structures (i.e., the posterior capsule) can limit forward movement of the tibia.

Lachman's Versus Anterior Drawer Test

- 🕮 Lachman's test may be more difficult for clinicians to perform but tends to be more sensitive.
- In the anterior drawer test, the knee is positioned so that the hamstrings have a mechanical advantage. Increased hamstring activity can inhibit tibial translation, causing a false negative test.
- A torn meniscus can act as a block to tibial motion, again causing a false negative while performing the anterior drawer test.

Pivot Shift Test

- Tests for anterolateral rotary instability.
- High specificity for ACL injury
- This maneuver reproduces the inciting event that occurs when knees give way. The patient often recognizes this as a buckling of the knee.
- The tibia is internally rotated while a valgus stress and axial load is applied.
- Begin in full knee extension and gently flex the knee.
- Anterolateral subluxation of the lateral tibial plateau indicates a positive test.
- The knee may be tested in full extension and at approximately 25 degrees of flexion to relax the fibers of the posterior joint capsules.
- 5 mm of motion is considered a Grade I tear.
- More difficult to perform than Lachman's or the anterior drawer test.

Tests for the Posterior Cruciate Ligament (PCL)

Posterior Drawer Test (Figure 4–114)

- The posterior drawer test evaluates the integrity of the PCL.
- The patient and examiner are in the same positions as for the anterior drawer test.
- The examiner pushes the tibia away from themselves.
- A positive test occurs when the tibia slides backward on the femur without a distinct endpoint. Some posterior movement is normal; however, a distinct endpoint should be encountered.

FIGURE 4–114 Positive posterior drawer sign: Torn PCL. PCL, posterior cruciate ligament.
Source: Courtesy of JFK Johnson Rehabilitation Institute, 2000.

Sag Test

- This is also a test for the integrity of the PCL.
- The patient is supine with the knee flexed to 90 degrees with the foot on the exam table.
- The test is positive if the tibia is displaced posteriorly. Comparison should be made with the contralateral side.
- The test can also be done with the patient supine with the hip and knee flexed to 90 degrees and the heel held in the examiner's hand.

Tests for Collateral Ligament Testing

Collateral Ligament Testing (Figure 4–115)

- This tests the integrity of the MCL and LCL by imposing valgus and varus stresses, respectively.
- With the patient supine, the examiner tucks the patient's ankle under the examiner's arm. The thumbs of both hands palpate the medial and lateral joint lines.
- If a **valgus stress** is applied and the medial joint line gaps, the MCL may be damaged **(Figure 4–115A)**.
- If a **varus stress** is applied and the lateral joint line gaps, the LCL may be damaged **(Figure 4–115B)**.
- The knee may be tested in full extension and at approximately 25 degrees of flexion to relax the fibers of the posterior joint capsules.
- 5 mm of motion is considered a Grade I tear.
- 5 to 10 mm is considered a Grade II tear.
- >10 mm is considered a complete interstitial tear.
- Be aware that muscle guarding may cause a false negative test

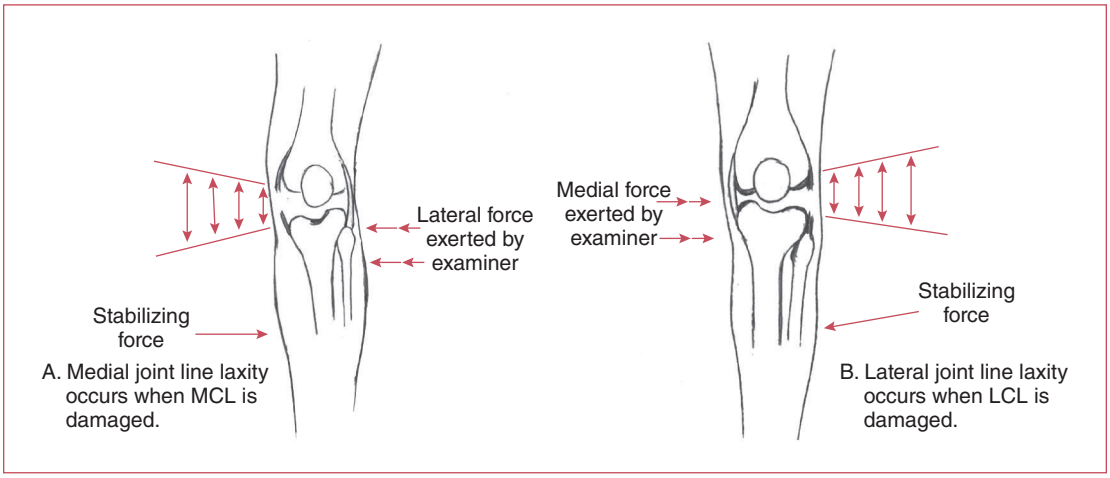

FIGURE 4–115 Collateral ligament testing. (A) Valgus stress test for integrity of the MCL. (B) Varus stress test for integrity of the LCL.
LCL, lateral (radial) collateral ligament; MCL, medial (ulnar) collateral ligament.

KNEE DISORDERS

MENISCAL INJURIES

General

- Meniscal tears are caused by shearing forces from loading and rotational forces on the knee.
- Medial meniscal injuries are associated with cutting maneuvers. They occur with tibial rotation while the knee is partially flexed during weight bearing (closed kinetic chain):
  - Medial meniscal injuries are common in sports such as football, basketball, and soccer.
- Lateral meniscal injuries typically occur during squatting. Full flexion with rotation is the usual mechanism (e.g., wrestling).

Clinical Features

- Acute meniscal tears:
  - Acute tears are often associated with a pop after a specific inciting incident.
  - Obstructive meniscal tears may cause true locking or buckling, with the torn meniscal tissue blocking joint ROM.

  - Posterior horn tears of the medial meniscus are common and occur with valgus and external rotation.
  - Effusions may occur within 24 hours.
  - Patients frequently complain of knee stiffness.
- Degenerative tears:
  - These typically occur with an insidious onset but may involve minimal trauma.
  - They usually occur in patients >40 years old and are often asymptomatic.
  - Locking episodes may be minimal.
- Physical exam:
  - ROM is decreased:
    - Effusion will limit flexion.
    - Obstructive meniscal tear can also limit ROM.
  - Tenderness:
    - Medial joint line tenderness is suggestive of medial meniscal injury.
    - Lateral joint line tenderness indicates lateral meniscal damage.

PROVOCATIVE TESTS

- Apley's grind (see **Figure 4–108**) and McMurray's tests (see **Figure 4–107**)

Imaging

- MRI is the gold standard in diagnosing meniscal tears:
  - Sagittal views will best show the anterior and posterior meniscal horns.
  - Coronal views are the best views for the meniscal body.
  - Tears appear as a line of increased signal extending from articular surfaces.
  - Asymptomatic, degenerative meniscal tears are found in up to 60% of people over age 50.
- Arthrograms are less expensive than MRI but more invasive because they require injections of dye into the joint to assess meniscal integrity.

🕮 Treatment

- Physical therapy has been shown to be effective as initial treatment for nonobstructive meniscal tears.
- Surgical resection is often required with obstructive meniscal injuries to the inner two-thirds of the meniscus because of the area's avascularity and resultant poor tissue healing.
  - If the meniscus is resected, the patient is generally weight bearing as tolerated in 1 to 2 days.
- Injuries to the outer one-third of the meniscus are usually repaired due to better vascular supply.
  - If the meniscus is repaired, generally the patient is non-weight bearing for 4 to 6 weeks. Strengthening proceeds at that time.

ANTERIOR CRUCIATE LIGAMENT INJURIES

🕮 General

- The ACL **(Figure 4–116)** is the most commonly injured knee ligament in athletics (football, soccer, basketball, downhill skiing).
- The mechanism of injury is usually cutting, deceleration, and hyperextension of the knee:
  - Noncontact injuries are most common.
  - Contact injuries may often involve other structures:
    - >50% of ACL tears occur with meniscal tears.
    - The terrible triad (also known as O'Donoghue's triad) involves

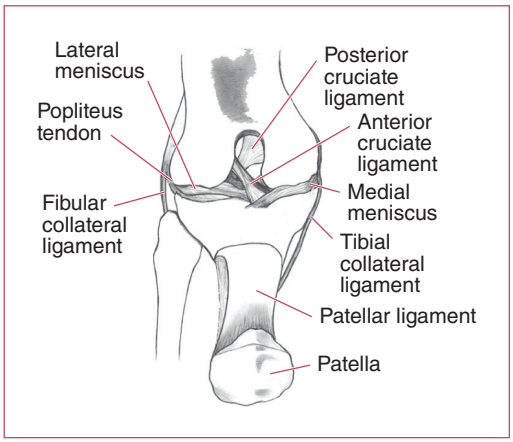

FIGURE 4–116 Anatomy of the ACL. ACL, anterior cruciate ligament.

simultaneous injury to the ACL, MCL, and medial meniscus because of the attachment of the MCL to the medial meniscus.
  - This injury occurs when a valgus force is applied to a flexed and rotated knee.

Clinical Features

- History:
  - There is a sudden pop and anterior knee pain with posterior lateral joint line pain.
  - Instability of the knee is common.
  - Early swelling: Within 24 hours, a significant effusion will be present. Severe effusion in the 2 to 12 hours following injury is the most sensitive marker for acute ACL injury.
- Physical exam:
  - An effusion is noted on clinical inspection.
  - Tenderness is variable and associated with meniscal tears and avulsion fracture.
  - The anterior drawer test (see **Figure 4–113**) may be positive or yield a false negative.
  - 🕮 Lachman's test (see **Figure 4–112**) may be positive but can yield a false negative in approximately 10% of cases. It is examiner-dependent and also influenced by muscle guarding.

Grading Criteria for Lachman Testing Classification

TRANSLATION GRADE	DEFINITION
I	<5 mm translation
II	5–10 mm translation
III	>10 mm translation
ENDPOINT GRADE	**DEFINITION**
A	Firm, sudden endpoint to passive anterior translation of tibia on fixed femur
B	Absent, ill-defined, or softened endpoint to passive anterior translation of tibia on a fixed femur

Source: From Mulligan EP, McGuffie DQ, Coyner K, et al. The reliability and diagnostic accuracy of assessing the translation endpoint during the Lachman test. *Int J Sports Phys Ther*. 2015;10(1):52–61.

  - Grading based on perceived anterior translation of the tibia on physical exam and perception of endpoint firmness:
    - Grade 1: 3 to 5 mm of translation
    - Grade 2: 5 to 10 mm translation, likely reflecting partial tear
    - Grade 3: >10 mm translation, likely reflecting complete tear`

Imaging and Testing

- X-rays may show an avulsion fracture of either the tibial insertion of the ACL or the proximal lateral capsular margin of the tibia (Segond's fracture pathognomonic for ACL tear).
- Arthrocentesis can be performed to relieve pressure and pain and will generally return blood or a sanguineous fluid in ACL tears.
- MRI is considered to be 85% to 90% accurate, showing rupture or partial tearing of the ACL.
- Arthroscopy is close to 100% accurate.

🕮 Treatment

- Initially partial weight bearing, ice, and compression are used while evaluation is ongoing.
- Nonoperative management in patients who are low demand based on activity as well as lower laxity such as Grade 1, as well as those who already have significant loss of meniscal integrity.
- Reconstruction is undertaken in younger, higher-level patients, especially Grade 3 as well as prior reconstruction failures.

- Postoperative course:
  - Partial weight bearing is maintained initially.
  - ROM is instituted to regain flexion over the first 2 weeks.
  - Progression to closed chain kinetics is then undertaken.
  - Avoid open chain exercises, especially those that are performed near full extension.
  - Resistive exercises performed between 0 and 45 degrees flexion are avoided during the first 3 to 6 months.
  - Lenox Hill derotation orthosis is used to control knee axial rotation as well as AP and medial–lateral control.
  - Sports-specific exercises may be started in 6 to 12 weeks.
  - Complete rehabilitation in 6 months to 1 year is the goal with maximum ROM, strength, and agility.

POSTERIOR CRUCIATE LIGAMENT INJURIES

General

- The most frequent cause of PCL injury is impact to the front of the tibia with the knee flexed (e.g., dashboard injury). The tibia is forced backward in relation to the femur, causing injury to the PCL.
- In athletics, hyperflexion is a common mechanism of PCL injury.
- PCL injuries are much less common than ACL injuries.

Clinical Features

- History:
  - The initial injury may or may not be associated with a pop.
  - There may be minimal swelling initially, increasing over 24 hours.
  - The ability to fully extend may be impaired.
  - The patient may be able to bear weight without pain.
- On physical exam:
  - An effusion may be present.
  - Popliteal tenderness is a common finding in the acute phase.
  - Posterior drawer test (see **Figure 4–114**) and sag tests may be positive (quadriceps spasms may cause a false negative).

Imaging

- X-rays may show an avulsion of the tibia.
- MRI is less accurate than for ACL tears.
- Arthroscopy has a higher accuracy than MRI.

Treatment

- Surgical repair is indicated if the ligament is avulsed with a tibial fragment.
- There is some controversy over surgical repair of an otherwise isolated PCL tear.
- Rehabilitation: Early prone passive mobilization with progressive weight bearing and quadriceps strengthening.

MEDIAL COLLATERAL LIGAMENT TEARS

General

- The MCL is the most commonly injured ligament of the knee.
- MCL injuries are common in football and skiing.
- Impact force to the lateral knee is often the mechanism of injury.
- However, MCL tears may occur without a direct blow. A sustained valgus force may also cause the injury.

Clinical Features

- History:
  - Often, there is a lateral blow (valgus stress) to the knee and a pop.
  - Medial knee pain is often immediately present.
  - Complete tears may allow walking and running after initial pain.
  - The knee becomes stiff in several hours.
- Physical exam:
  - Medial swelling and tenderness may be present and variable.
  - Minimal effusion may be present.
  - Medial instability on valgus stress testing is present.
  - Gap of 5 to 8 mm compared with the opposite side may indicate a complete tear.
  - Instability in slight flexion of 30 degrees is specific for MCL injury, whereas instability in full extension may indicate injury to the MCL and the posterior capsule.
  - The terrible triad of MCL tear, ACL tear, and medial meniscal tear (O'Donoghue's triad) is a possible complication and requires evaluation.

Imaging

- Radiographs may reveal an epiphyseal fracture.
- MRI is useful to delineate the MCL tear and also to investigate associated injuries (i.e., to the ACL and medial meniscus).
- US may visualize a tear of the MCL.

Treatment

- Isolated MCL tears may be treated conservatively.
- The knee can be braced.
- Rehabilitation focuses on strengthening and stability.
- Epiphyseal fractures may be present with or without medial collateral tears.
- Tears with concomitant injuries may require surgical intervention.

LATERAL COLLATERAL LIGAMENT TEARS

- Isolated LCL injuries are rare—evaluate for concurrent posterolateral corner knee instability.
- LCL tears usually are the result of knee dislocations.
- Consideration should be made of associated vascular, cruciate, and peroneal (fibular) nerve injuries.

ILIOTIBIAL BAND SYNDROME

General

- The ITB slides over the lateral femoral condyle with the knee in flexion and extension.
- The ITB extends from the TFL distally in the lateral leg to insert on Gerdy's tubercle on the lateral tibia.
- Dysfunction caused by inflexibility of the ITB and adductor/abductor muscle imbalances.

Clinical Features

- The patient presents with pain over the lateral femoral condyle and/or Gerdy's tubercle, which is exacerbated by walking or jogging. Symptoms improve with running.
- The patient adapts by externally rotating the hip, internally rotating the lower leg, and pronating the foot.
- ITB tightness is evaluated by the Ober test (for description of the Ober test, refer to the Lower Extremities: The Hip and Pelvis section).
- Knee pain associated with ITB tightness is further assessed by the following: The patient extends the knee and at approximately 30 degrees experiences pain over the lateral femoral condyle as the ITB crosses the bony prominence.

Imaging

- Generally not necessary for diagnosis. Radiographs are useful to evaluate possible avulsion fracture of Gerdy's tubercle if it is suspected.

🕮 Treatment

- Stretching the ITB, hip flexors, and gluteus maximus is central to rehabilitation.
- Strengthening the hip abductors, gluteus maximus, and TFL is also important.
- Orthotics may be helpful and foot overpronation must be corrected.
- Injection at the lateral femoral condyle may be necessary in resistant cases.

PATELLA-RELATED INJURIES

The stability of the patella is dependent upon three main characteristics:

1. Depth of the intercondylar groove
2. Proper contour of the patella
3. Adequate muscular control
   - The normal patellar motion is vertical.
   - At full extension, the applied force of the quadriceps approximating the patella to the condyles is reduced.
   - Patellofemoral weight bearing increases with knee flexion:
     - Walking: 0.5 times body weight
     - Ascending or descending stairs: 3.3 times body weight
     - Squatting: 6.0 times body weight
   - In hyperextension, there is a tendency for the patella to separate from the femur. The lateral lip of the patellar surface of the femur acts to prevent subluxation.

RECURRENT PATELLAR SUBLUXATION

General

- If a congenital malformation causes a less prominent lateral lip or a more prominent medial lip, the patella may dislocate laterally in full extension.
- Increased genu valgum laterally displaces the patella.
- Increased genu varum medially displaces the patella.
- Excessive genu recurvatum elongates the patellofemoral structures, causing loss of patella condylar contact.
- Vastus medialis weakness allows lateral displacement.
- Tibial external torsion can cause lateral displacement.
- A shallow lateral femoral condyle can cause lateral displacement.
- A laterally attached infrapatellar tendon on the tubercle can cause lateral displacement.

Clinical Features

- The patella may be displaced medially or laterally in the acute phase.
- The knee tends to buckle after a subluxation.
- Pain and tenderness are present in the peripatellar region.
- An effusion may be present.
- Wasting of the vastus medialis may be present.
- Full extension may be impaired.
- The patella will often reset at 25-to 30-degree flexion.

Imaging

- Radiographs
  - The AP view visualizes the patellar position over the sulcus.
  - The lateral view ascertains the patellar height and is done at 45-degree knee flexion and in full extension.
  - The sunrise (tunnel) view ascertains the patellofemoral articulation and femoral condyle height.

Treatment

- See the following for treatment of patellofemoral pain and overload syndrome.

PATELLOFEMORAL PAIN SYNDROME

🕮 General

- Also known as runner's knee or biker's knee
- With regard to bicycling, bicycle fit, recent change in equipment, and training distance and intensity are factors to consider.
- This may be the most common cause of anterior knee pain syndrome.
- It is an overuse injury caused by repeated microtrauma, leading to peripatellar synovitis.
- The predisposing conditions noted previously in recurrent patellar subluxation apply for this syndrome. They are both patellar tracking problems.

Clinical Features

- The syndrome presents as anterior knee pain of acute or insidious onset.
- An effusion may be present.
- Crepitus may be present on ROM.
- Ascending or descending stairs tends to aggravate the condition.
- Patellar compression produces the pain in the patellofemoral compartment.
- Examination may reveal a high-riding, laterally shifted patella (**patella alta**). This condition is due to vastus lateralis tightness and relative medial weakness, causing tracking dysfunction.
- A low patella (**patella baja**) is less common and may indicate quadriceps rupture.
- Examination of the knee in the last 30-degree extension is important.
- A tight lateral retinaculum and/or vastus medialis oblique (VMO) dysplasia can lead to lateral patellar shift or shear stress, resulting in cartilage damage.
- Rotation of the patella also indicates evidence of muscle imbalances:
  - Patellar internal rotation is given the term *squinting patella*.
  - Patellar external rotation is given the term *frog's eye patella*.
- Tight hip flexors can alter gait and cause symptoms:
  - Check with the Thomas test (see Lower Extremities: The Hip and Pelvis section).
- Measure Q angle. Normal: Females should be approximately 18 degrees; males should be approximately 13 degrees **(see Figure 4–100)**:
  - 🕮 Factors that increase Q angle: Internal torsion of the femur, lateral insertion of the infrapatellar tendon on the tibia, genu valgum.
- Tight abductors can also alter gait:
  - Check with the Ober test (see Lower Extremities: The Hip and Pelvis section).
- Tight hamstrings can increase patellofemoral loading.
- Check with the straight leg raise test.

Imaging

- Radiographs
  - The AP view visualizes the patellar position over the sulcus.
  - The lateral view ascertains the patellar height and is done at 45-degree knee flexion and full extension.
  - The sunrise (tunnel) view ascertains the patellofemoral articulation and femoral condyle height.
- MRI
  - MRI is *not* often used to assess patellofemoral pain. Articular degeneration may be seen (see chondromalacia patella).
- CT
  - CT is an alternative. It is also useful if a growth plate injury is suspected in pediatric patients.
  - It can evaluate the stage of patellar subluxation present in the last 15-degree flexion that plain films may not reveal.
  - CT can also reveal and delineate tumors.

- Bone scan
  - Bone scan is useful to evaluate symptoms present for <4 months of uncertain diagnosis.
  - It is a useful technique to detect intra-articular processes: Osteochondritis dissecans, osteomyelitis, or tumor.

Treatment

Conservative:

- The vast majority is successfully treated by conservative means.
- Controlling symptoms is of primary concern achieved by:
  - Decreasing pain
  - Increasing strength
  - Increasing ROM
- Activity modification
  - For active individuals: Reduce the pace of activities that increase patellofemoral stress (climbing, jumping, squatting). Also correct any mechanics that may be contributing to the issue.
  - For inactive individuals: Institute baseline exercises and progress incrementally as tolerated.
- Proper bicycle fit
  - Adjust saddle position. Having the saddle too far forward or too low will stress the anterior knee but too high may lead to pain in the posterior knee.
  - Minimizing hyperflexion of the cyclist knee in the force generation phase of pedaling is key to preventing anterior pain from patellofemoral pain syndrome (PFPS) or patellar tendinitis.
  - Adjust rotation of the cleats. Anterior knee is stressed with internally rotated cleats, whereas the medial knee is stressed with externally rotated cleats.
- Ice 10 to 15 minutes four to six times per day is used in the acute phase. This may be required before and after activity in the subacute phase.
- NSAIDs should be used acutely and then tapered off as pain decreases.
- Therapeutic exercise
  - Quadriceps strengthening, especially the VMO, is important:
    - Short arc quadriceps activities (0–15 degrees) are used to strengthen the VMO.
    - Isotonic quadriceps strengthening with eccentric loading occurs in a nonpainful ROM.
    - Isometric quadriceps contractions (quad sets) are used.
    - Straight leg raising for iliopsoas strengthening occurs.
    - Stretching of the hamstrings, ITB, adductors, and vastus lateralis is also important to balance forces.
    - Proprioceptive exercises are used after initial strengthening and stretching.
    - Increase activity when ROM is full and pain-free and strength is 80% of normal.
- Patellar taping
  - Useful adjunct in therapy.
  - The patella is taped in position to allow pain-reduced ROM:
    - Provides proprioceptive feedback to alter patellar tracking
    - Also helps to balance contractile forces between the vastus lateralis and medialis
- Patellar bracing (knee sleeve with patellar cutout or strap)
  - Provides proprioceptive feedback
  - May prevent reinjury
  - Can allow the patient to resume activity

Surgical:

- Surgery is rarely needed. Consider surgical referral if:
  - Conservative measures fail after 4 to 6 months
  - An obvious surgical lesion exists
- Surgery may involve:
  - Lateral release of the knee capsule and retinaculum
  - Patellar realignment
  - Patellar tendon transfer
  - Patellectomy

CHONDROMALACIA PATELLA

General

- Chondromalacia patella is characterized by softening of the patellar articular cartilage followed by cartilage degeneration.
- MRI is only moderately accurate in diagnosing chondromalacia. Diagnostic arthroscopy is considered the gold standard for diagnosis, and the cartilage appears roughened or fibrillated. These are findings noted in PFPS.
- Chronic patellofemoral overload and tracking dysfunctions may predispose to this condition. Therefore, the functional and anatomic conditions noted in recurrent patellar subluxation and patellofemoral overload syndrome apply.
- Degradation of the articular cartilage of the patella may also result from infection, trauma, or autoimmune processes.

Clinical Features

- The presentation of chondromalacia patella is clinically the same as for PFPS, which was described previously.

Imaging

- The imaging workup of chondromalacia patella is the same as for PFPS as previously noted.
- MRI may detect defects in the articular cartilage of the patella, which is the distinguishing characteristic of this disorder.

Treatment

- Because abnormal patellar mechanics are often the cause of chondromalacia patella, treatment will follow the treatment already described for PFPS.

PLICA SYNDROME

General

- The synovial lining of the knee extends from the infrapatellar fat pad medially around the femoral condyles and under the quadriceps tendon above the patella and lateral to the lateral retinaculum.
- Plicae are redundant folds of the synovial lining of the knee. They are common and generally asymptomatic.
- Plica syndrome refers to a pathologic condition in which the plica becomes inflamed. It is a diagnosis of exclusion.
- Plica syndrome can occur in the mediopatellar, infrapatellar, suprapatellar, or lateral regions.

Clinical Features

- As noted earlier, plicae are typically asymptomatic.
- The patient may present with anterior knee pain of insidious onset that can gradually increase with prolonged knee flexion or sitting but is made worse by standing and extension.
- After direct trauma, such as ACL tears or meniscal tears, the plica can become inflamed and symptomatic.
- The knee may give the sensation of buckling if the plica is trapped between the patella and medial condyle.
- Careful palpation with knee flexion and extension may detect a plica.
- Snapping with knee extension may occur if the plica is fibrosed.

Imaging and Testing

- Because plica syndrome often accompanies patellofemoral dysfunction, the imaging workup correlates what is typically done for patellofemoral pain.
- MRI is useful in evaluation of the plica as well as other potential causes of knee pain.
- Arthrogram and arthroscopy can also be used.

Treatment

- 🕮 The treatment would follow that of PFPS as previously outlined.
- Surgical excision can be done if conservative measures fail.

PATELLAR TENDONITIS (JUMPER'S KNEE)

General

- 🕮 Patellar tendonitis is often associated with microtears of the patellar tendon.
- 🕮 Patellar tendonitis is an overuse syndrome of the patellofemoral extensor unit. It is associated with repetitive, high quadriceps loading: Jumping, squatting, kneeling, and climbing stairs.
- 🕮 The most common site of involvement is the inferior pole of the patella.
- The superior pole of the patella or the insertion over the tibial tubercle may also be sites of involvement.

Clinical Features

- The patient presents with pain on high-impact activity, such as jumping.
- The pain may diminish during the activity session and become more apparent afterward.
- There will be tenderness on palpation over the patellar tendon in addition to the inferior or superior pole of the patella.

Imaging

- X-ray of the knee to evaluate bony anatomy.
- MRI of the knee can assess the integrity of the patellar tendon.

Treatment

- Treatment follows the guidelines for PFPS as previously described.

🕮 OSTEOCHONDRITIS DISSECANS

General

- This condition is thought to be secondary to repetitive small stresses to subchondral bone that disrupt blood supply to that area of bone.
- This process is in a localized segmental area of AVN at the end of a long bone. The cause of osteochondritis dissecans in children and adults remains unclear.
- This results in the formation of dead subchondral bone covered with articular hyaline cartilage.
- The overlying cartilage degenerates around the defect and an entire piece may detach from the rest of the bone, entering the joint space as a loose body.
- The usual area of involvement in the knee joint is the medial femoral condyle.
- 🕮 Other areas that may get involved include other areas of the distal femur, patella, elbow, talus, and distal humerus.
- The condition most commonly affects adolescents.
- It is thought to be secondary to repetitive small stresses to subchondral bone that disrupt the blood supply to that area of bone, but there is no clear evidence to support this.

Clinical Features

- The patient may experience gradual onset of joint pain and irritation, synovial effusion, and a buckling sensation.
- Walking with the foot rotated outward may relieve pain.
- To palpate the medial femoral condyle, have the knee flexed 90 degrees with pressure directed medial to the inferior pole of the patella.
- The presence of a loose body may cause locking.

Imaging

- The area of involvement may be visible on radiographs. MRI will better define the lesion area.

Treatment

- The healing of the defect may occur if the diagnosis is made before the fragment separates:
  - The knee must be placed at rest and protected from weight bearing.
- If healing does not occur or if the fragment becomes detached, the lesion will require surgical excision.

🕮 POPLITEUS TENDONITIS

General

- The popliteus muscle arises from the lateral face of the lateral femoral condyle and inserts into the triangular area in the posterior tibia.
- Its main function is internal rotation of the tibia (laterally rotates femur on the tibia = medially rotating tibia with respect to the femur, depending on which bone is fixed).
- The popliteus muscle also assists in unlocking the knee by laterally rotating the femur.
- With the ACL, it limits anterior translation of the *femur*.
- Inflammation of the popliteus tendon can be due to acute or chronic overload.

Clinical Features

- Lateral knee pain typically during downhill activities and with excessive pronation
- Stress to the popliteus is caused by forward femoral displacement, such as running downhill.
- Point tenderness anterior to the LCL
- Pain with legs in figure-of-four or cross-legged position

Treatment

- Avoid downhill running until symptoms resolve.
- Rest, ice, compression, NSAIDs
- May require arch supports or medial heel wedges

LOWER EXTREMITIES: THE LOWER LEG

FUNCTIONAL ANATOMY

Muscle Groups

The three muscle groups found in the lower leg are defined by their anatomic relation to the tibia: Anterior, lateral, and posterior.

Anterior Muscle Group (Figure 4–117)

- This group is composed of ankle dorsiflexors, toe extensors, and foot evertors/inverters.
- **Ankle dorsiflexors and foot invertors:**

ANKLE DORSIFLEXORS AND FOOT INVERTORS	SPINAL INNERVATION	PERIPHERAL INNERVATION
Tibialis anterior (TA)	L4, L5	Deep peroneal (fibular) nerve
Extensor hallucis longus (EHL)—great toe extensor	L4, L5	Deep peroneal (fibular) nerve

- **Ankle dorsiflexors and foot evertors:**

ANKLE DORSIFLEXORS AND FOOT EVERTORS	SPINAL INNERVATION	PERIPHERAL INNERVATION
Extensor digitorum longus—toe extensor	L4, L5	Deep peroneal (fibular) nerve
Peroneus (fibularis) tertius	L4, L5	Deep peroneal (fibular) nerve

Lateral Muscle Group (Figure 4–118)

- This group functions as foot evertors and weak plantar flexors.
- **Foot evertors and weak plantar flexors:**

FOOT EVERTORS AND WEAK PLANTAR FLEXORS	SPINAL INNERVATION	PERIPHERAL INNERVATION
Peroneus (fibularis) brevis	L5, S1	Superficial peroneal (fibular) nerve
Peroneus (fibularis) longus	L5, S1	Superficial peroneal (fibular) nerve

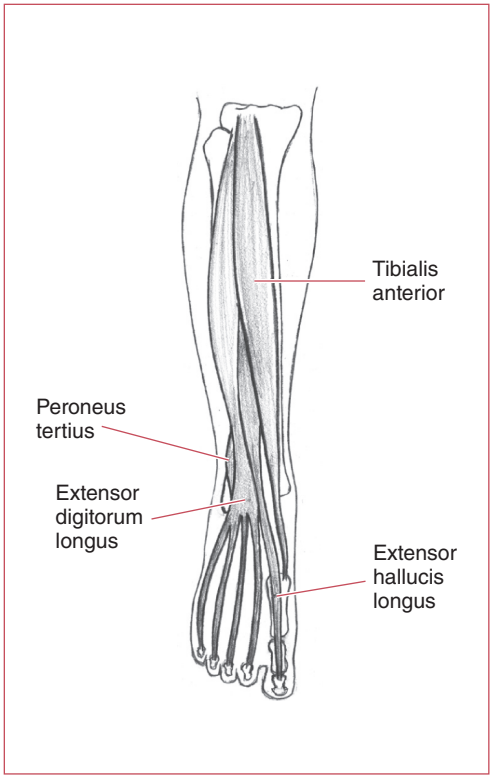

FIGURE 4–117 Anterior muscle group of the lower leg.
*The terms peroneus and fibularis are used interchangeably. The word "peroneus" has been kept in the figure; however, please note that peroneus = fibularis.

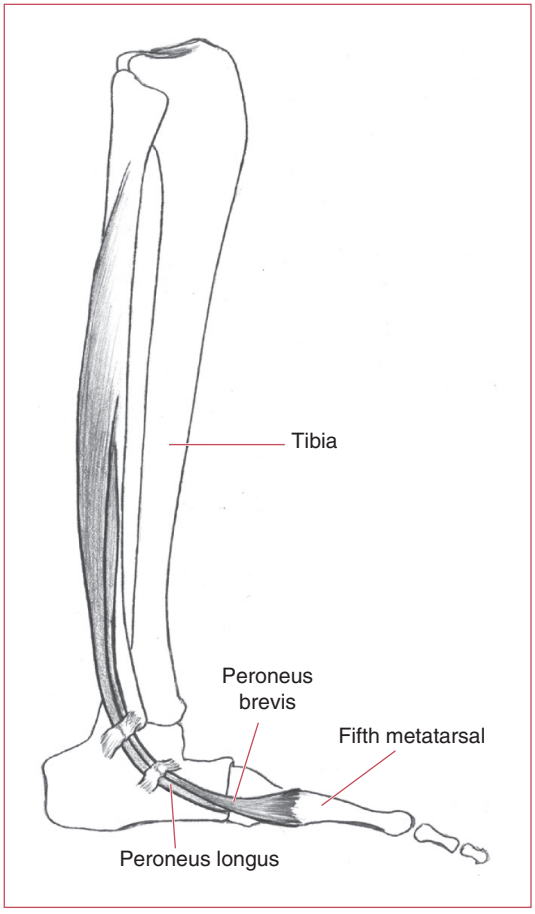

FIGURE 4–118 Lateral muscle group of the lower leg.

Posterior Muscle Group (Figure 4–119)

POSTERIOR MUSCLE GROUP	SPINAL INNERVATION	PERIPHERAL INNERVATION	ACTION
SUPERFICIAL POSTERIOR COMPARTMENT			
Gastrocnemius	L5, S1, S2	Tibial nerve	Plantar flexor
Soleus	L5, S1, S2	Tibial nerve	Plantar flexor
Plantaris	L5, S1, S2	Tibial nerve	Weak plantar flexor
DEEP POSTERIOR COMPARTMENT			
Flexor digitorum longus	L5, S1, S2	Tibial nerve	Flexion lateral four toes, inversion, plantar flexor
Tibialis posterior	L5, S1, S2	Tibial nerve	Inversion and plantar flexor
Popliteus	L5, S1, S2	Tibial nerve	Internal rotation of the leg on the femur
Flexor hallucis longus	S2, S3	Tibial nerve	

Compartments (Figure 4–120)

THERE ARE FOUR COMPARTMENTS OF THE LOWER LEG

1. Anterior compartment
   The **anterior compartment** comprises the tibialis anterior (TA), extensor digitorum longus, extensor hallucis longus, and peroneus (fibularis) tertius muscles; the anterior tibial artery and vein; and the deep peroneal (fibular) nerve.
2. Lateral compartment
   The **lateral compartment** comprises the peroneus (fibularis) longus and brevis muscles, the superficial peroneal (fibular) nerve, and the common peroneal (fibular) nerve dividing into the superficial and deep branches.
3. Deep posterior compartment
   The **deep posterior compartment** comprises the flexor digitorum longus, flexor hallucis longus (FHL), tibialis posterior, and popliteus muscles; the posterior tibial artery and vein and peroneal (fibular) artery and vein; and the tibial nerve.

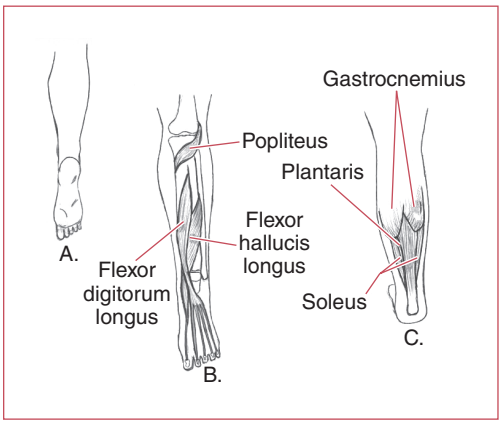

FIGURE 4–119 Posterior muscles of the leg. (A) Posterior plantar flexion. (B) Posterior plantar flexion, intermediate deep layer of posterior muscle group. (C) Posterior aspect, superficial layer of posterior muscle group.

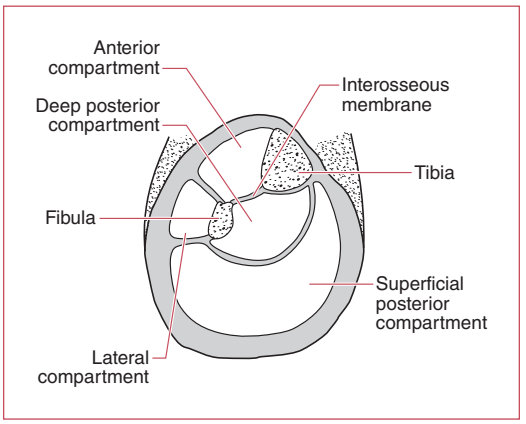

FIGURE 4–120 Lower leg in cross-section. (Note fascial compartments.)

4. Superficial posterior compartment
   📖 The superficial posterior compartment comprises the gastrocnemius, soleus, and plantaris muscles. The gastrocnemius and soleus complex is also known as the triceps surae.

DISORDERS OF THE LOWER LEG

CHRONIC EXERTIONAL COMPARTMENT SYNDROME

General

- This condition is marked by chronically raised intracompartmental pressure, most commonly affecting the anterior compartment.
- Pressure increases have been measured during and after exercise.
- Nerve impingement can result from these high intracompartmental pressures, either during or after exercise.
- Tissue ischemia may develop due to restricted arterial inflow, obstruction of microcirculation, or arteriolar/venous collapse.
- The pain associated with this syndrome may be explained by:
  - Sensory receptor stimulation in the fascia or periosteum caused by high pressure.
  - Release of biochemical factors caused by reduced blood flow.
- Lower leg pain may also be caused by tibial stress fractures or periostitis. These syndromes can present concurrently with chronic exertional compartment syndrome (CECS).

Clinical Features

- 📖 In CECS, pain will typically increase with exercise and progress as the activity increases in intensity.
- Pain will usually diminish after the activity is stopped. Occasionally, it may persist if the exercise session was particularly demanding.
- Neurologic involvement can cause weakness and numbness:
  - Anterior CECS, affecting the deep peroneal (fibular) nerve, can cause ankle dorsiflexion weakness and numbness over the first web space of the dorsum of the foot.
  - Lateral CECS, affecting the superficial peroneal (fibular) nerve, can cause foot eversion weakness and numbness of the dorsal foot and anterolateral distal shin.
  - Deep posterior CECS can cause weakness with plantar flexion and numbness of the plantar foot (tibial nerve).
  - On palpation, the compartment may be firm, and after exercise, deep palpation may be painful.
  - Fascial hernias may be palpable.
  - Rarely, distal pulses can be diminished.

Imaging and Testing

- 📖 Compartment pressure measurement using manometric technique is the gold standard to evaluate CECS.
  - Elevated pressures should coincide exactly with reproduction of the patient's pain syndrome.
  - Pressures are measured pre- and post-exercise. A delay in return to pre-exercise pressure levels of 6 to 30 minutes is required for a positive test.
- Slit/wick catheter technique: Inserting a catheter into the compartment and monitoring the pressure via a transducer connected to a pressure amplifier and recorder.
- MRI has been found useful by some practitioners to diagnose CECS.
- Plain films, CT, bone scan, and MRI help to investigate other sources of leg pain.
- EMG and NCS may help diagnose neuropathy as well as unrelated sources of symptoms.
- EMG/NCS pre- and post-exercise have been tried as a diagnostic tool with limited results.

Treatment

- At present, the most effective treatment for CECS is fasciotomy.
- Conservative management strategies often involve physical therapy, analgesic medications, modifying pain-inducing activities, and reducing biomechanical stresses (through gait and footwear adjustments).

ACUTE COMPARTMENT SYNDROME

General

- Acute and significantly increased intracompartmental pressure results in ischemia of muscle and nerve tissues and can lead to necrosis.
- In an acute compartment syndrome (ACS), the intracompartmental tissue pressure acutely becomes elevated and produces a secondary elevation in venous pressure that obstructs venous outflow. An escalating cycle of continued increase in intracompartmental tissue pressure occurs.
- Necrosis of muscle and nerve tissue can develop in as little as 4 to 8 hours. Thus, recognition and treatment of ACS constitutes a surgical emergency.
- ACS most commonly follows trauma in association with fractures of the long bones of the leg or forearm.
- The volar aspect of the forearm and the anterior compartment of the leg (shin area) are the most commonly affected. The tibia is the most commonly associated bone.
- Cases of ACS without a fracture were at significantly greater risk for delayed diagnosis and treatment.
- ACS from nontraumatic causes are less frequent, including but not limited to ischemia-reperfusion injury, s/p revascularization procedures, vascular disease, thrombosis, conditions that decrease serum osmolarity, certain animal bites, or after prolonged limb compression (e.g., following severe drug or alcohol intoxication or poor positioning during surgery).
- If untreated, tissue necrosis can develop with secondary muscle paralysis, muscle contractures, and sensory impairment called Volkmann's ischemic necrosis. It results in claw hand or foot caused by contractures of the ischemic muscle.

Clinical Features

- Intractable pain and sometimes sensory hypesthesia distal to the involved compartment (usually dorsal foot and median distribution in the hand) are the most important early symptoms.
- The characteristics are pain, paresthesia, and paralysis (3 Ps).
- Exam
  - Remove cast or dressing if present.
  - The most important physical sign is *extreme pain* on stretching the long muscles passing through a compartment (plantar flexion of toes or full extension of fingers will stretch muscles of the forearm or shin).
  - Compartment manometric measurement (with catheter insertion) of suspicious increases in compartment syndrome is mandatory.
  - Note: Pulses are usually completely normal in ACS because the intracompartmental pressure rarely exceeds systolic or mean arterial pressure levels.

Imaging and Testing

- Manometric techniques (handheld manometry or needle manometry) are available to measure compartment pressure directly.
- Slit catheter technique—inserting a catheter into the compartment and monitoring the pressure via a transducer connected to a pressure amplifier and recorder
- General guidelines indicate the presence of a compartment syndrome when the absolute compartment pressure (ACP) is >30 mmHg or pressure differential (diastolic blood pressure [BP] – ACP) is ≤30 mmHg (Cone & Inaba, 2017).

Treatment

- In ACS, muscle necrosis may develop in 4 to 8 hours.
- When diagnosis is made, emergent surgical fasciotomy of the compartment is essential. The wound is left open with delayed closure or skin grafting performed after the edema subsides.
- Without immediate treatment, ACS may result in permanent loss of function and tissue damage, including muscle necrosis and nerve damage. Fingers and toes often claw and have little motion.

MEDIAL TIBIAL STRESS SYNDROME

General

- Commonly known as shin splints, this condition occurs from overuse or repetitive stress, resulting in chronic traction on the periosteum at the periosteal–fascial junction.
- It is a common cause of exercise-induced leg pain.
- The periosteum may become detached from the bone due to ballistic overload.
- A fibrofatty filling may occur at the site of the defect.
- The attachment of the soleus muscle along the medial tibia may be the most likely site of avulsion.
- In some cases, the other deep flexors of the foot are involved.
- 🕮 The main predisposing factor to shin splints is hyperpronation.

Clinical Features

- The patient presents with the gradual onset of pain along the posteromedial border of the tibia.
- 🕮 Pain may improve with exercise but worsens *after* the completion of the activity and can last until the next morning.
- There will be tenderness on palpation along the medial tibial border along the entire length or for as little length as 2 to 4 cm.
- There may be a history of repetitive running on hard surfaces and inappropriate warm-up, inappropriate footwear, or recent change in footwear.
- There may be a history of excessive use of the foot flexors, as in jumping.

Imaging

- Plain films will be normal. Bone scan may reveal areas of uptake along the medial tibial border in the third phase. MRI will help rule out a tibial stress fracture.

Treatment

- Relative rest is the first point in management of medial tibial stress syndrome (MTSS). Decreasing the volume of the inciting activity or avoidance of the activity entirely should be considered.
- When pain is present at rest or with normal walking, crutch walking is indicated.
- Icing and stretching are also done initially.
- Return to activity should be gradual and should occur when the patient is pain-free for several days. Soft, level surfaces should be used initially.
- A comprehensive history of the patient's exercise routine should be obtained, including running mileage, changes in training routine and intensity, and footwear. Common training errors include increasing training regimens "too much, too fast" and extensive running on hard or uneven surfaces.
- Training should start at 50% of preinjury level for intensity and distance.
- Hyperpronation is also a significant risk factor for MTSS. Orthotics are useful to correct overpronation or forefoot varus.
- Fasciotomy of the posteromedial fascia may be required in resistant cases but is rarely needed.

STRESS FRACTURES

General

- Tibial stress fractures are the most commonly occurring stress fractures in running sports.
- See Table 4–2 for incidence of lower extremity stress fractures in runners.
- They are a common cause of lower leg pain in athletes (25% incidence rate).
- Stress fractures occur when repetitive loading causes irreversible bone deformation.
- Microfractures develop, and with continued overuse are not compensated by normal bony growth and turnover. As a result, they can coalesce and propagate through the bone to become a symptomatic stress fracture.

TABLE 4–2 Incidence of Lower Extremity Stress Fractures in Runners

BONE	INCIDENCE (%)
Femoral neck	7
Femoral upper shaft	5
Distal femoral shaft	2
Fibula	24
Tibia 1. Metaphysis 2. Upper shaft 3. Midshaft 4. Distal shaft	34 7 12 4 11
Metatarsals 1. Second (most common stress fracture) 2. Third 3. Other	20 11 7 2
Sesamoid Os calcis, talus, navicular	2

Source: Adapted from DeLisa JA, Gans BA, eds. *Rehabilitation Medicine: Principles and Practice*. 2nd ed. Lippincott Williams & Wilkins; 1993, with permission.

- Low bone mineral density (BMD) poses a higher risk for stress fractures. Causes of low BMD include:
  - Females with late onset of menses
  - Individuals with low body weight (<75% ideal body weight)
  - Poor nutrition correlates with lower calcium intake
  - Tobacco and excessive alcohol use
  - Female athlete triad
- Biomechanical conditions can lead to a higher risk of stress fracture.
  - Overpronation places higher stress on the fibula and tibia (e.g., genu valgum and a wide gait pattern may cause overpronation).
  - Leg length discrepancies, muscle imbalances, lack of flexibility, and malalignment factors can also place high forces on the lower leg.
  - Poor conditioning.
- Extrinsic factors can lead to stress fractures.
  - The classic training errors of increasing intensity and length of activity abruptly ("too much, too soon") or causing excessive fatigue increases the chances of stress fractures.
  - Running on hard surfaces or using worn-out shoes may also be a cause.

Clinical Features

- Pain worsens with increased activity.
- The pain will typically be localized to the fracture site.

- Night pain may occur.
- On palpation, there is tenderness localized to the fracture site.
- There may be swelling and redness at the fracture site.

Imaging

- X-rays are the first-line diagnostic imaging but are rarely abnormal initially.
  - Stress fractures may not be seen for 2 to 3 weeks after symptoms develop.
  - Periosteal thickening appears first, followed by cortical lucency.
  - The linear stress fracture appears as a lucency within a thickened area of cortical hyperostosis during healing.
- MRI
  - MRI is the most sensitive and specific, becoming the first choice of test in investigating stress fractures.
  - MRI has comparable sensitivity to bone scan with better specificity.
  - MRI can also visualize the fracture and differentiate between fracture and other lesions.
- Bone scan
  - Bone scans can be used when radiographs are negative and a stress fracture is still suspected.
  - Bone scans are very sensitive (close to 100%) but low in specificity.
  - A positive scan will yield focal increased uptake in the third phase.
  - Tumors, osteomyelitis, bony infarct, and bony dysplasia can also cause localized increased uptake.
- CT
  - CT scan can reveal fracture lines not visualized with plain films.
  - CT can also differentiate other lesions that cause increased uptake on bone scan.

Treatment

- Treatment depends on the severity of the fracture, its site, and its duration of symptoms.
- Stress fractures of the first to fourth metatarsal shafts typically will heal with rest alone and do not require immobilization.
- If there is pain on normal ambulation, the patient should be non-weight bearing for 7 to 10 days.
- Those without pain on normal ambulation should avoid aggravating the injury (i.e., relative rest).
- NSAIDs and ice may be used initially to control pain.
- Muscle strengthening and stretching can begin immediately.
- Nonimpact activities such as cycling and swimming can be used to maintain cardiac fitness.
- At least 1 to 2 weeks of pain-free normal ambulation should be achieved before returning to impact activity.
- Impact activity is started cautiously at low intensity for short periods (10–15 minutes) in the first few days and increased incrementally as tolerated.
- Orthotics are a useful adjunct to treatment and may decrease impact force and help correct excessive supination or pronation.
- Softer running surfaces should be considered.
- Evaluation of exercise routine should be performed.
- Calcium intake should be 1,500 mg daily with 400 to 800 IU of vitamin D.

LOWER EXTREMITY: THE ANKLE AND FOOT

FUNCTIONAL ANATOMY

Ankle Mortise (Figure 4–121)

- The distal tibia forms the medial malleolus and part of the superior articular surface of the ankle mortise joint.

- The distal fibula forms the lateral malleolus and part of the superior articular surface of the ankle mortise joint.
  - The lateral malleolus extends more distally than the medial malleolus, making it very important in ankle stabilization.
- Talus comprises four parts:
  - Body, neck, head, and dome
  - It has a fragile blood supply, which can lead to complications of healing.

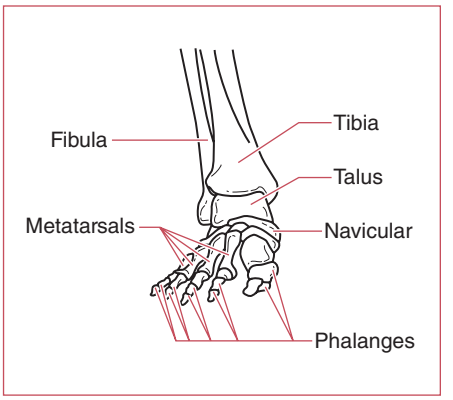

FIGURE 4–121 Ankle mortise.

Bones of the Foot (Figure 4–122)

- **Seven tarsals**
  - Talus
  - Calcaneus
  - Navicular
  - Cuboid
  - Three cuneiforms (medial, intermediate, and lateral)
- **Five metatarsals**
- **Fourteen phalanges**
  - Proximal, middle, distal
  - The great toe has just proximal and distal phalanges.
- **Two sesamoid bones**
  - Located on the plantar surface of the head of the first metatarsal

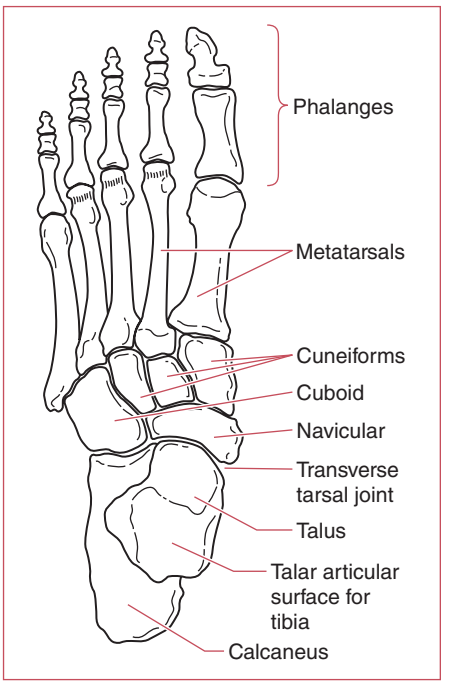

FIGURE 4–122 Bones of the foot and ankle: Dorsal view.

Ligaments of the Ankle (Figure 4–123)

- 🕮 **Lateral aspect**
  - Anterior talofibular ligament (ATFL)
    - Primary lateral ankle ligament stabilizer
    - Most commonly injured ligament
  - Calcaneofibular ligament (CFL)
  - Posterior talofibular ligament (PTFL)
- **Medial aspect**
  - The deltoid ligament stabilizes the medial ankle joint and is stronger than the lateral ligaments. It consists of deep and superficial components.
    - *Deep* (more important in maintaining stabilization):
      - Anterior tibiotalar
      - Posterior tibiotalar
    - *Superficial:*
      - Tibionavicular
      - Tibiocalcaneal
  - The deltoid ligament maintains proximity of the medial malleolus and talus, preserving the medial longitudinal arch of the foot.
    - Spring ligament (plantar calcaneonavicular ligament)
    - Medial and posterior talocalcaneal ligaments
- **Anterior aspect:** Syndesmotic ligaments
  - Comprises four ligaments that maintain the integrity of the distal tibia and fibula as well as resisting any forces that would separate the two bones
  - Ligaments
    1. Anterior tibiofibular ligament
    2. Posterior tibiofibular ligament
    3. Transverse tibiofibular ligament
    4. Interosseous ligament

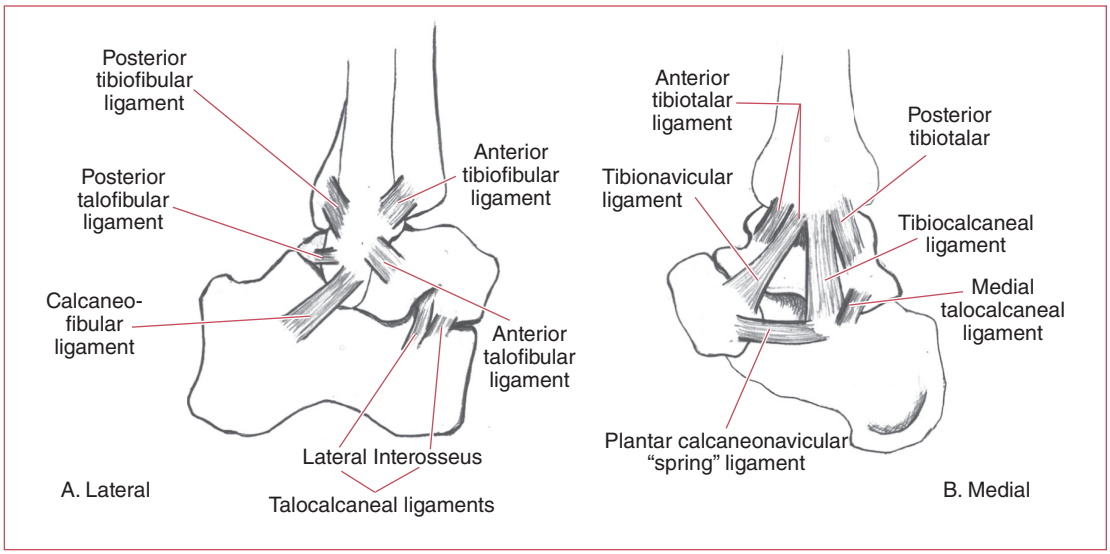

FIGURE 4–123 Ligaments of the ankle and subtalar joint. (A) Lateral view. (B) Medial view.

Ligaments of the Foot

- **Lisfranc ligament**—connects the second metatarsal head to the first cuneiform
- **Transverse metatarsal ligament**

FOOT AND ANKLE ROM

- **Ankle**
  - Dorsiflexion: 20 degrees
  - Plantar flexion: 50 degrees
- **Foot**
  - Subtalar joint
    - Inversion: 5 degrees
    - Eversion: 5 degrees
  - Forefoot
    - Abduction: 10 degrees
    - Adduction: 20 degrees
  - First metatarsal phalangeal (MTP)
    - Flexion: 45 degrees
    - Extension: 80 degrees

	ANTERIOR MUSCLE GROUP	
	INVERSION	**EVERSION**
Dorsiflexion	Tibialis anterior Extensor hallucis longus	Extensor digitorum longus Peroneus (fibularis) tertius
	POSTERIOR MUSCLE GROUP	**LATERAL MUSCLE GROUP**
	INVERSION	**EVERSION**
Plantar flexion	Tibialis posterior Flexor digitorum longus Flexor hallucis longus Plantaris Gastrocnemius Soleus	Peroneus (fibularis) longus Peroneus (fibularis) brevis

Foot Motion

TOE FLEXORS	SPINAL INNERVATION	PERIPHERAL INNERVATION
Flexor digitorum longus	S2, S3	Tibial nerve
Flexor digitorum brevis	S2, S3	Medial plantar nerve (tibial nerve)
Flexor hallucis longus	S2, S3	Tibial nerve
Flexor hallucis brevis	S2, S3	Medial plantar nerve (tibial nerve)
Quadratus plantae	S2, S3	Lateral plantar nerve (tibial nerve)
Interossei	S2, S3	Lateral plantar nerve (tibial nerve)
Flexor digiti minimi brevis	S2, S3	Lateral plantar nerve (tibial nerve)
Lumbricals First lumbrical Second, third, and fourth lumbricals	 L4, L5 S2, S3	 Medial plantar nerve (tibial nerve) Lateral plantar nerve (tibial nerve)

TOE EXTENSORS	SPINAL INNERVATION	PERIPHERAL INNERVATION
Extensor digitorum longus	L5, S1	Deep peroneal (fibular) nerve
Extensor digitorum brevis	L5, S1	Deep peroneal (fibular) nerve
Extensor hallucis longus	S1, S2	Deep peroneal (fibular) nerve
Lumbricals First lumbrical Second, third, and fourth lumbricals	 L4, L5 S2, S3	 Medial plantar nerve (tibial nerve) Lateral plantar nerve (tibial nerve)

TOE ABDUCTORS	SPINAL INNERVATION	PERIPHERAL INNERVATION
Abductor hallucis	S2, S3	Medial plantar nerve (tibial nerve)
Abductor digiti minimi	S2, S3	Lateral plantar nerve (tibial nerve)
Dorsal interossei	S2, S3	Lateral plantar nerve (tibial nerve)

TOE ADDUCTORS	SPINAL INNERVATION	PERIPHERAL INNERVATION
Adductors hallucis	S2, S3	Lateral plantar nerve (tibial nerve)
Plantar interossei	S2, S3	Lateral plantar nerve (tibial nerve)

DISORDERS OF THE ANKLE

LATERAL ANKLE DISORDERS

🕮 Lateral Ankle Sprains (Figure 4–124)

General

- Most common ankle sprain, accounting for up to 85% of all ankle sprains.

Mechanism of Injury

- Inversion of a plantar-flexed foot places the foot in the most vulnerable position to cause ligamentous injury.
- History of "rolling over" the ankle

FIGURE 4–124 Lateral ankle sprain.

Injured Ligaments

- ATFL
  - Most commonly injured ligament in lateral ankle sprains
- CFL
  - Second most commonly injured ligament
  - Stabilizes the ankle during inversion
- PTFL
  - Last ligament to be injured

Clinical Features

ANKLE SPRAIN GRADING

- Grade 1 (Mild):
  - Partial tear of the ATFL
  - CFL and PTFL are intact
  - Mild swelling with point tenderness at the lateral aspect of the ankle
  - No instability
  - Stress tests
    - Anterior drawer: Negative
    - Talar tilt: Negative
- Grade 2 (Moderate):
  - Complete tear of the ATFL
  - Partial tear of the CFL
  - Diffuse swelling and ecchymosis
  - Stress tests
    - Anterior drawer: Positive
    - Large anterior shift of the ankle or palpable clunk
    - Talar tilt: Negative
- Grade 3 (Severe):
  - Complete tear of the ATFL and CFL
  - Stress tests
    - Anterior drawer: Positive
    - Talar tilt: Positive
      - Inverting the talus on the tibia looking for a clinical asymmetry in comparison
- Dislocation
  - The majority of ankle fractures occur with a dislocation (fracture-dislocation).
  - Complete tears of the ATFL, CFL, and PTFL

PROVOCATIVE TESTS (FIGURE 4–125)

- **Anterior drawer test (Figure 4–125A)**
  - Tests the integrity of the ATFL
- **Talar tilt test/inversion stress test (Figure 4–125B)**
  - Tests the integrity of the CFL and ATFL
  - Inversion stress applied when ankle is plantar flexed

Imaging

- X-rays with AP, lateral, and oblique views
- Stress views: Anterior drawer and talar tilt
- Anterior drawer may show a >5 mm displacement.

- Talar tilt may show a 5- to 10-degree difference as compared with the contralateral extremity.
- Tilting more than 30 degrees may indicate that all three lateral ligaments are ruptured.
- US may demonstrate tears of the ATFL and CFL.

A

B

FIGURE 4–125 Stress tests for ligament stability. (A) Anterior drawer sign. (B) Inversion test (talar tilt).

Treatment

- Grades 1 and 2:
  - Acute
    - Rest, ice, compression, elevation (RICE), NSAIDs, analgesics, immobilization
    - Early mobilization
  - Conservative: Rehabilitation
    - Physical therapy: ROM, strengthening, proprioceptive exercises, taping, and bracing
    - Modalities: Moist heat, warm whirlpool, contrast baths, US, short wave diathermy
- Grade 3:
  - Controversial: Conservative versus surgical
    - Six-month trial of rehabilitation and bracing
    - Ligament repair, tenodesis of the peroneus (fibularis) brevis
    - If the patient is a high-performance athlete, and conservative treatment fails (i.e., patient has persistent critical instability), then surgical reconstruction of torn ligaments may be considered as early as 3 months postinjury.
    - 🕮 Indications for surgery include large bony avulsions, severe ligamentous damage on the medial and the lateral sides of the ankle, and severe recurrent injuries.
- Grade 1: Weight bearing as tolerated. Grade 2, Grade 3: Non-weight bearing.

Peroneal (Fibular) Tendon Injury

General (Figure 4–126)

- Peroneus (fibularis) longus inserts on the base of the first metatarsal.
- Peroneus (fibularis) brevis inserts on the base of the fifth metatarsal.
- These muscles plantar flex the ankle and evert the foot.

Mechanism of Injury

- Tenosynovitis or rupture
  - Repetitive forceful eversion causing inflammation or degeneration of the tendon or synovium along its course, behind the lateral malleolus to its insertion point
- Subluxation or dislocation
  - A sudden dorsiflexion of the ankle with the foot can cause a subluxation or dislocation of the peroneal (fibular) tendon. This insult is commonly a skiing injury.

Clinical Features

- Painful swelling in the lateral retromalleolar area along the course of the peroneal (fibular) tendons

- Sudden weakness with the inability to actively evert the foot if a subluxation or dislocation is suspected
- A popping sensation in the lateral aspect of the ankle

PROVOCATIVE TEST

- Pain with resisted dorsiflexion and eversion

Imaging

- None needed initially. MRI if indicated.
- US may help visualize peroneal (fibular) tendon subluxation over the lateral malleolus when the ankle is dorsiflexed and everted.

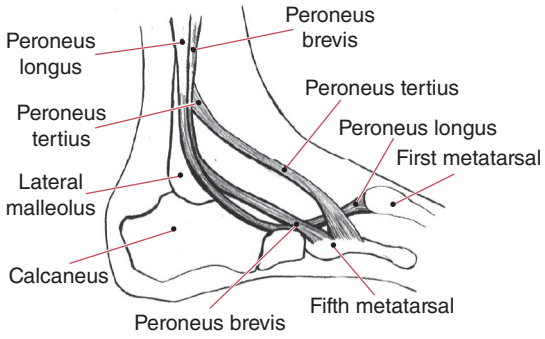

FIGURE 4–126 Anatomy of peroneus (fibularis) longus, brevis, and tertius.
*The terms peroneus and fibularis are used interchangeably. The word "peroneus" has been kept in the figure. However, please note that peroneus = fibularis.

Treatment

- Tenosynovitis: Same treatment as for a lateral ankle sprain.
- Rupture/subluxation/dislocation → orthopedic evaluation—4 to 6 weeks of immobilization in a plantar-flexed position.

MEDIAL ANKLE DISORDERS

Medial Ankle Sprain: Deltoid Ligament

General

- Rare ankle injury associated with a 5% occurrence rate
- 🕮 Medial ankle ligaments are stronger than lateral ankle ligaments.
- Deltoid ligament is one ligament comprising four parts **(Figure 4–127)**:
  - Anterior tibiotalar ligament
  - Posterior tibiotalar ligament
  - Tibionavicular ligament
  - Tibiocalcaneal ligament
  - – Function to stabilize the ankle during eversion
  - – The deltoid ligament is the most prominent medial ankle stabilizer.
    - Especially during inversion

Mechanism of Injury

- Foot caught in a pronated, everted position with internal rotation of the upper body. An example is when the foot strikes the ground instead of the ball in soccer or an extra point in football.
- Pure eversion is rare.

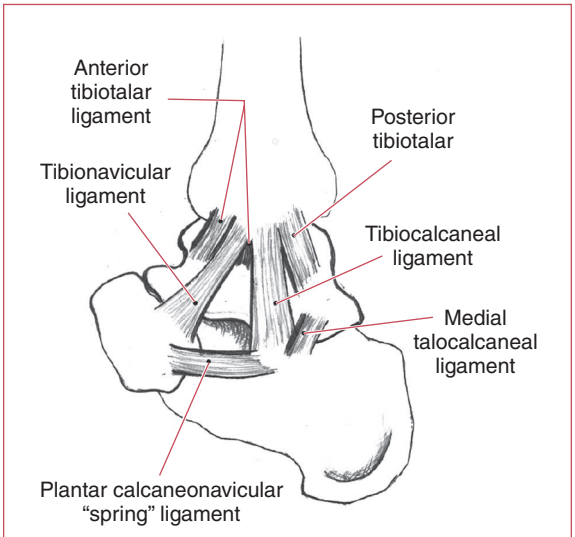

FIGURE 4–127 The deltoid ligament.

Grading Scale

- Grade 1: Stretch
- Grade 2: Stretch partial tear
- Grade 3: Full tear

Complications

- Syndesmosis ankle injuries and Maisonneuve fractures (see Tibiofibular Syndesmosis Injury section)

Clinical Features

- Medial foot swelling and ecchymosis and pain on eversion

PROVOCATIVE TEST

- Negative anterior drawer test
- Positive eversion test (eversion of ankle stresses the deltoid ligament)

Imaging

- X-rays with AP, lateral, and oblique views
- MRI if indicated

Treatment

- Same as lateral ankle injuries
- Immobilization for 4 to 6 weeks
- Molded shoe orthosis for 4 to 6 months
- Surgery if conservative measures fail

Tibialis Posterior Tendon Injury

General (Figure 4–128)

- Anatomy
  - Origin: Interosseous membrane and the posterior surface of the tibia and fibula
  - Insertion: Tuberosity of the navicular, cuboid, and base of the second to fourth metatarsals
- Function: Plantar flexes the ankle and inverts the foot
  - Maintains the medial longitudinal arch

Mechanism of Injury

- Tenosynovitis or tendon rupture
  - Repetitive forceful inversion causing inflammation or degeneration of the tendon or synovium along its course
  - 5% to 10% associated with an accessory navicular bone

Clinical Features

- Insidious onset of posteromedial ankle pain increased by activity
- Medial hindfoot swelling

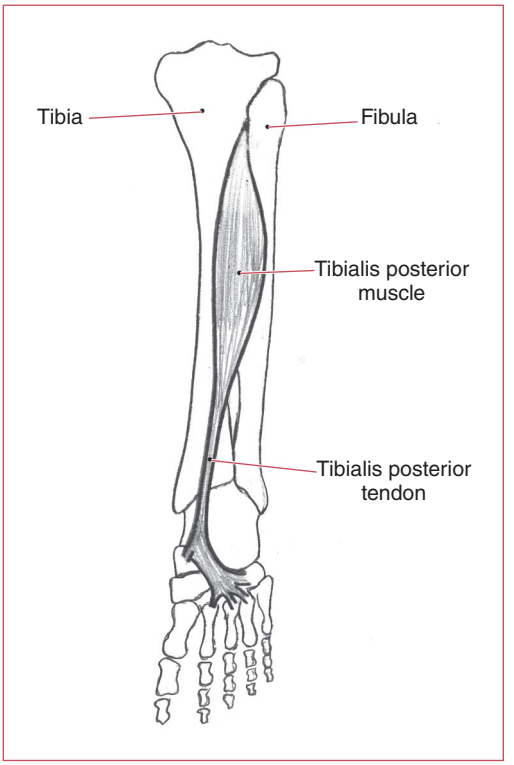

FIGURE 4–128 Tibialis posterior muscle and tendon (posterior view).

- Increased pain with push-off
- Weakness with inversion and plantar flexion
- The classic "too many toes" sign is seen with a tibialis posterior tendon rupture (**Figure 4–129**).
  - When viewing the patient's feet from behind, more toes are visible on the affected side secondary to collapse of the medial longitudinal arch.
- Malicious malalignment syndrome is a combination of a broad pelvis, increased femoral anteversion, squinting patellae, excessive Q angle, and excess pronation of the foot. Hyperpronation can lead to tibialis posterior pain.

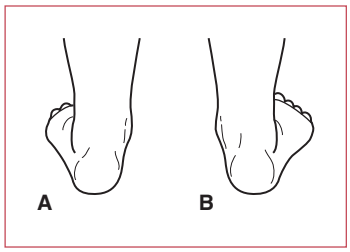

FIGURE 4–129 (A) Normal. (B) Positive "too many toes" sign.

Imaging

- None needed. MRI if indicated.

Treatment

- *Acute:* Same as lateral ankle sprains
- *Conservative:* Rehabilitation, orthotics to address pronation
- *Surgical:* Tendon transfer, excising the accessory navicular

POSTERIOR ANKLE DISORDERS

Achilles Tendon Disorders

General (Figure 4–130)

- Gastrocsoleus complex function
  - Plantar flexes the ankle
  - Supinates the foot

Mechanism of Injury

- Achilles tendonitis
  - Repetitive eccentric overload causing inflammation and microtears of the tendon
- Achilles tendon rupture
  - **Inflammatory:** Inflammation and degeneration causing a series of microtears or breakdown in the collagen fibers, which can lead to tendon rupture
  - 🕮 **Vascular:** Inadequate vascularization 2 to 6 cm proximal to the insertion of the tendon
  - 🕮 **Mechanical:** Sudden strong push-off (active plantar flexion); the tendon can similarly rupture with aggressive dorsiflexion as the gastrocsoleus is contracted.

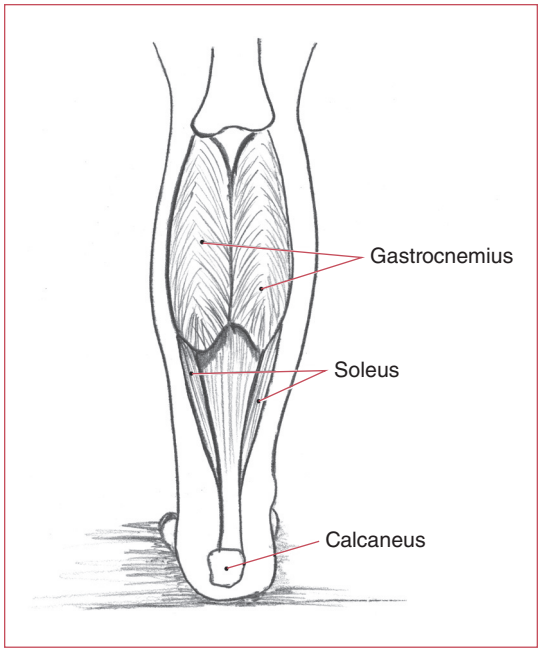

FIGURE 4–130 Anatomy of the gastrocnemius and soleus muscles.

Risk Factors for Achilles Tendonitis

- Training errors: Most common risk factor
  - Sharp increase in mileage or intensity
  - Change in recent footwear
- Anatomic causes: Hyperpronation, tight hamstrings and heel cords (Achilles tendons), pes cavus, and genu varum

- Increased age leads to decreased tendon flexibility and decreased tensile strength.

Clinical Features

- Tendonitis:
  - Posterior ankle pain, swelling
  - Pain elicited on push-off
- Achilles tendon rupture:
  - 🕮 Sudden audible snap with immediate swelling, ecchymosis, and weakness in plantar flexion.
  - With a negative Thompson's test (**Figure 4–131A**), squeezing the calf should elicit plantar flexion of the foot.
  - A positive Thompson's test (**Figure 4–131B**) is indicative of a ruptured Achilles tendon, and the foot is unable to plantar flex secondary to the tendon rupture.

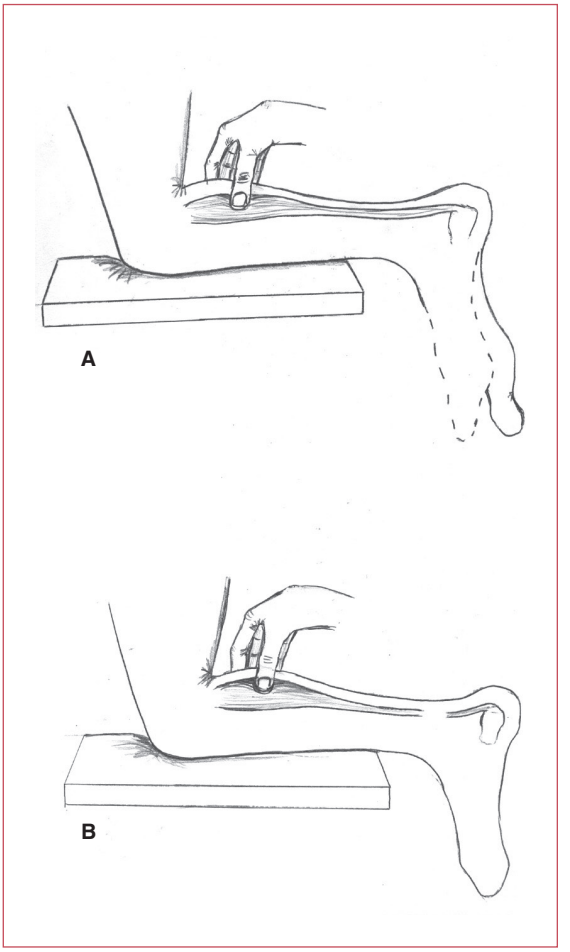

FIGURE 4–131 Thompson's test. (A) Negative. (B) Positive.

Imaging

- None needed for Achilles tendonitis. MRI is helpful if the diagnosis is in question.
- US and MRI may be helpful in differentiating partial from complete tear.

Treatment

- 🕮 **Achilles tendonitis:**
  - Relative rest, ice, anti-inflammatory medications
  - Rehabilitation: Short-term immobilization (splinting or bracing), stretching and strengthening, heel lifts
  - Do not inject corticosteroids into the Achilles tendon, as it may cause tendon rupture.
    - 🕮 Complication of rupture: The area of hypovascularity 2 to 5 cm proximal to the tendon insertion is where most ruptures occur. Corticosteroids decrease the metabolic rate of the chondrocytes and fibrocytes, weakening the structural integrity of the tendon and articular cartilage.
- **Achilles tendon rupture:**
  - *Conservative:* Non-weight bearing with crutches and cast/controlled ankle movement (CAM) boot, followed by physical therapy after 2 weeks. Continued non-weight bearing using crutches and cast/CAM boot in a plantar-flexed position for a period of 8 to 12 weeks. The dorsiflexion is increased gradually so that by the end of 12 weeks, the patient is in a neutral position. Activity level and weight-bearing status are increased gradually (heel lifts may be used).
  - *Surgical:* Tendon repair. Patient is placed in a cast for 2 weeks and then put in a plantar flexion dial lock brace for 4 to 6 weeks, which is slowly brought to neutral. Activity level is increased gradually (heel lifts may be used). Surgical repair is usually reserved for active individuals.

Flexor Hallucis Longus Injury

General

- Also known as dancer's tendonitis
- Anatomy (**Figure 4–132**)
  - Origin: Distal fibula and interosseous membrane
  - Insertion: Base of the distal phalanx of the great toe
- Function
  - Flexes the great toe at all the joints
  - Plantar flexes the ankle

Mechanism of Injury

- Repetitive push-off maneuvers cause inflammation of the synovium or tendon as it courses in the groove of the sustentaculum tali and behind the lateral malleolus to its insertion.

Clinical Features

- Tenderness along the tendon at the posteromedial aspect of the great toe
- Decreased ability to flex the great toe
- Increased pain with active plantar flexion and passive dorsiflexion

Imaging

- None needed

Treatment

- Same as lateral ligament sprain

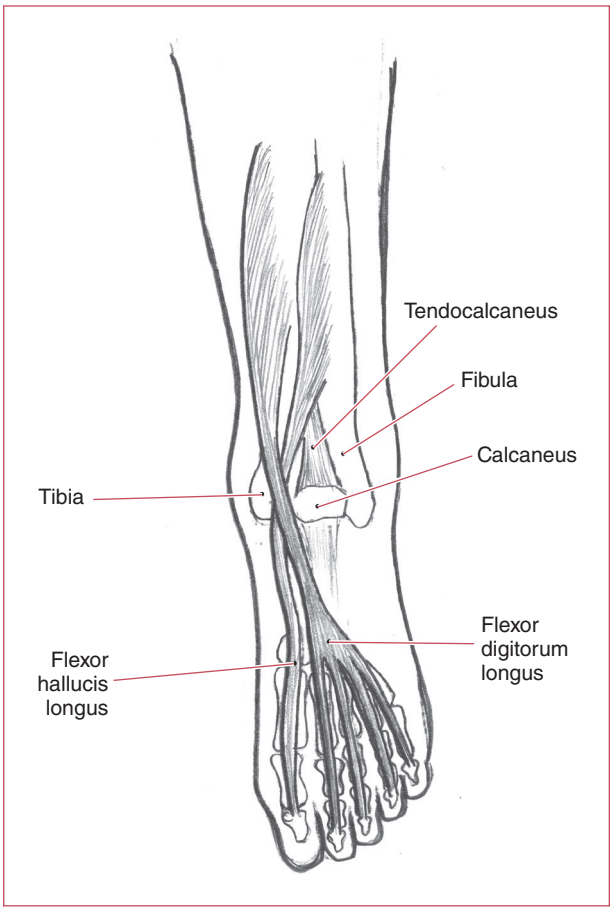

FIGURE 4–132 Anatomy of FHL (posterior view—plantar flexed). FHL, flexor hallucis longus.

BURSITIS: RETROCALCANEAL BONY EXOSTOSIS, CALCANEAL APOPHYSIS

Haglund Syndrome

General

- Retrocalcaneal pain caused by a triad of:
  - Retrocalcaneal bursitis
  - Achilles tendinitis/tendinopathy
  - Haglund deformity (i.e., posterosuperior calcaneal exostosis)
- Retrocalcaneal bursitis is a common cause of posterior ankle pain, especially in women, secondary to high heel shoes.
- Retrocalcaneal bursa anatomy (**Figure 4–133**): Inflammation of the bursae between the posterior superior portion of the calcaneus and the distal Achilles tendon, or a bursa between the skin and the Achilles tendon causing pain.

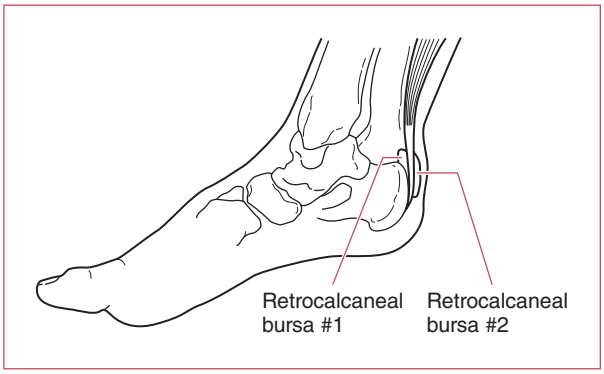

FIGURE 4–133 Retrocalcaneal bursa anatomy: Medial view.

Mechanism of Injury

- Repetitive pressure and shearing forces from an object (e.g., high heels or shoes with a hard heel counter), causing thickening and inflammation of the bursae, Achilles tendon, and enlargement of the posterior calcaneal region.
- Associations:
  - **Haglund's deformity** ("pump bump"): Retrocalcaneal exostosis (enlargement of the posterosuperior tuberosity).
  - **Sever's disease:** Calcaneal apophysitis: An independent area of ossification separated from the main bone at the cartilaginous plate. It occurs in the younger population, especially among female gymnasts from the stresses of the gastrocnemius pull.

Clinical Features

- Tenderness and swelling in the posterior aspect of the distal tendon/tendon insertion region
- Small soft tissue swelling at the Achilles insertion can be palpated on exam, "pump bumps."
- Similar types of pain can result from insertional Achilles tendonitis or retrocalcaneal bursitis.

Imaging

- None needed

Treatment

- Change or alter footwear
- Surgical excision of the bursae if conservative measures fail

ANTERIOR ANKLE DISORDERS

Tibiofibular Syndesmosis Injury

General

- Also known as a high ankle sprain
- Tibiofibular syndesmosis refers to the articulation of the tibia and fibula, which is held together by ligaments. Its function is to:
  - Maintain the integrity of the ankle mortise.
  - Resist forces that attempt to separate the tibia and fibula.
- Ligamentous anatomy **(Figure 4–134)**:
  - Anterior tibiofibular ligament
  - Posterior tibiofibular ligament
  - Interosseous ligament
  - Transverse tibiofibular ligament

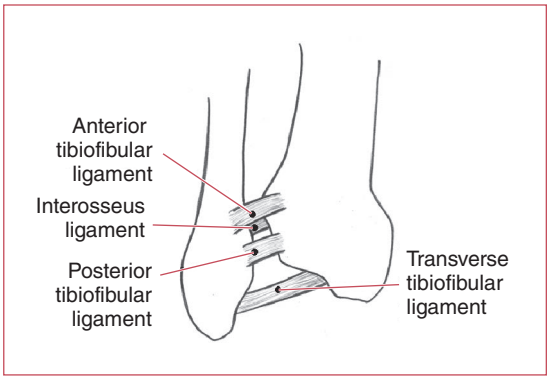

FIGURE 4–134 The syndesmotic ligaments of the ankle.

Mechanism of Injury

- Hyperdorsiflexion and forceful eversion of the ankle
- Direct blow to the foot with the ankle held in external rotation

Clinical Features

- Chronic in nature; pain and swelling on the anterior aspect just above the ankle

PROVOCATIVE TESTS

- **Squeeze test:**
  - Tenderness on compression of the distal tibia and fibula proximal to the injury

- **Stress test:**
  - With the knee held at 90 degrees and ankle neutral, the patient experiences pain when the examiner attempts forcefully to externally rotate the foot.

Imaging

- Plain films with AP, lateral, oblique, and mortise views. MRI, CT.
- Rule out widening between the distal tibia and fibula.
- Be sure to also obtain plain films of the proximal fibula to rule out the possibility of a Maisonneuve fracture **(Figure 4–135)**.
- 🕮 Maisonneuve fracture: Rupture of the anterior tibiofibular ligament extending through the interosseous membrane, which often results in a proximal fibula fracture

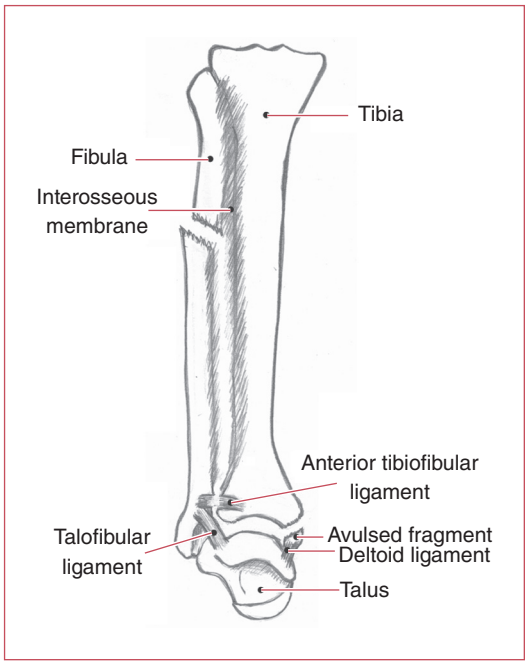

FIGURE 4–135 Maisonneuve fracture.

Treatment

- Conservative
- *Surgical:* Screw fixation to stabilize the ankle mortise

Sinus Tarsi Syndrome

General

- Talocalcaneal ligament sprain

Mechanism of Injury

- Excessive foot pronation causing adduction of the talus
- History of arthritis: RA, gout, and seronegative spondyloarthropathies
- History of prior ankle injury: Inversion sprain or fracture of the tibia, calcaneal, or talus

Clinical Features

- Pain on the anterolateral aspect of the foot and ankle in the area of the sinus tarsi **(Figure 4–136)**
- Diagnosis: Resolution of symptoms with injection of local anesthetic into the sinus tarsi

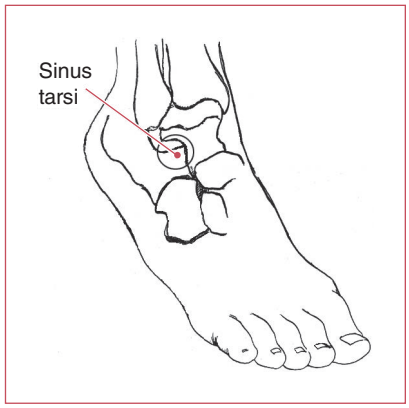

FIGURE 4–136 Sinus tarsi area.

Imaging

- None needed

Treatment

- *Conservative:* Same as lateral ankle sprain; corticosteroid injection
- *Surgical:* Decompression of the tunnel contents

Tibialis Anterior Tendon Injury

General

- The TA muscle dorsiflexes the ankle and inverts the foot.

Mechanism of Injury

- Tenosynovitis
  - Inflammation of the tendon or synovium as it courses under the superior retinaculum
- Tendon rupture
  - Degenerative process seen in the elderly
  - Eccentric overload

Clinical Features

- The TA can be palpated distally upon its insertion onto the medial aspect of the base of the first metatarsal and the first cuneiform bones (**Figure 4–137**) and proximally along the tendon to the muscle belly on the lateral side of the tibial shaft.
- Patient with a history of chronic ankle pain
- Painless foot slap that has gradually increased over time
- Increased tenderness and weakness with active dorsiflexion and passive plantar flexion
- Palpable defect may be noted over the anterior aspect of the ankle.

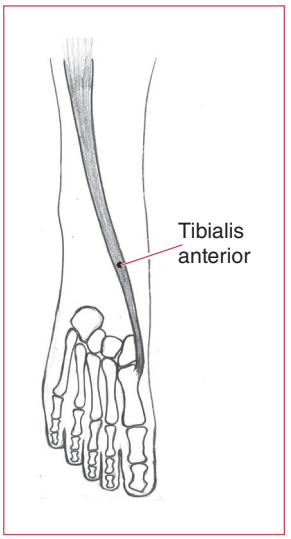

FIGURE 4–137 Palpation of the tibialis anterior tendon.

Imaging

- None needed. MRI if indicated.

Treatment

- Conservative versus surgical, depending on the patient's age and functional needs

Tibialis Anterior Spasticity (also see Spasticity Section in Chapter 12, Associated Topics in Physical Medicine and Rehabilitation)

General

- Hyperactivity of the TA contributes to inversion (varus) and supination positioning in equinovarus deformities

Clinical Features

- Commonly seen in patients with upper motor neuron lesions: Cerebral palsy, CVA, and traumatic brain injury (TBI). Any of these injuries may lead to spasticity of the TA.
- Equinovarus deformity is present on physical exam.
- Achilles tendon is frequently shortened, exacerbating the equinus posture.

Imaging

- Plain films—AP, lateral

Treatment

- *Conservative*
  - ROM, stretching, casting, bracing, chemoneurolysis, neurotoxin injections (botulinum toxin)
- *Surgical*
  - 🕮 SPLATT procedure: **SPL**it **A**nterior **T**ibial **T**endon transfer
- Technique: The TA tendon is split, and a portion of the tendon is transferred to the lateral foot. Half remains attached to its site of origin, while the distal end of the lateral half of the tendon is tunneled into the third cuneiform and cuboid bones.
- This provides an eversion force to counteract the dynamic varus deformity to provide a flat base for weight bearing.

- The SPLATT procedure is often done along with Achilles tendon lengthening to decrease plantar flexion.
- Overcorrection is a possible complication.

Talar Neck Fractures

General

- Etiology:
  - Shear force on the anterior lateral surface of the talus results in a shallow lesion.
  - Compressive force on the posterior medial surface of the talus results in a deep lesion.

Mechanism of Injury

- Eversion and dorsiflexion
- Inversion and plantar flexion

Hawkins' Classification

- Type I: Nondisplaced vertical fracture of the talar neck
- Type II: Displaced fracture of the talar neck of the subtalar joint with the ankle joint remaining intact
- Type III: Displaced fracture of the talar neck with dislocation of the body of the talus from the subtalar and ankle joints

Complications

- 🕮 AVN, most commonly of the talar body. The risk of AVN increases as the amount of displacement increases.
- Fracture of the talar dome may form a subchondral fragment that can detach and become displaced in the joint space.

Clinical Features

- Chronic ankle injury
- Small effusion that may be painful
- Decreased ROM

Imaging

- Plain films: Mortise view of the ankle; MRI

Treatment

- *Conservative:* Non-weight bearing
- *Surgical:* ORIF if indicated

FOOT DISORDERS

PLANTAR FASCIITIS (FIGURE 4–138)

General

- Medial plantar heel pain caused by inflammation of the plantar fascia
- Affects females > males
- Other associations: HLA-B27; seronegative spondyloarthropathy. Heel spurs may contribute to the etiology: 50% to 75% with heel spurs have plantar fasciitis.

Mechanism of Injury

- Increased tension on the plantar fascia leads to chronic inflammation, most commonly at its origin.
- Disorders causing tension include pes cavus (high arch), pes planus (flat foot), obesity, tight Achilles tendon, and bone spurs.

🕮 Clinical Features

- Tenderness is observed over the medial aspect of the heel at the origin of the plantar fascia and along the plantar arch.
- Pain can be elicited by hyperextension of the great toe with palpation along the plantar fascia.
- Pain is worse in the morning or at the start of weight-bearing activities (standing, walking after prolonged sitting) and decreases during activity.
- Tight Achilles tendon is frequently associated with plantar fasciitis.

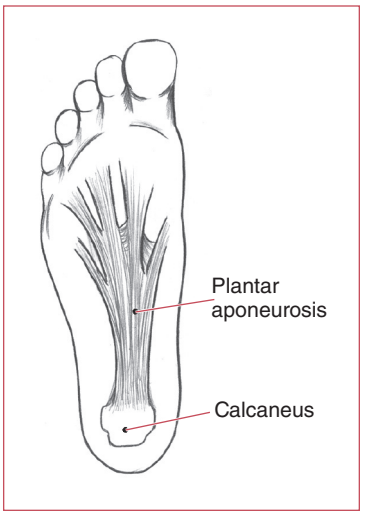

FIGURE 4–138 Plantar aponeurosis.

Imaging

- Plain films to assess for bony spur

Treatment

- *Conservative:* 90% to 95% effective and should be done for at least 6 months prior to considering surgery:
  - Modalities, NSAIDs
  - Orthotics; shoe modifications (heel pads, cushion, and lift)
  - Achilles tendon and plantar fascia stretching (negative plantar fascia stretch on a step, eccentric calf stretches, roll tennis ball under plantar surface)
  - 🕮 Injections: Do not inject anesthetic/corticosteroid into the subcutaneous tissue or fascial layer. Stay out of the superficial fat pad to avoid fat necrosis.
  - Nighttime dorsiflexion splints if other conservative measures fail
- *Surgical:* Plantar fascia release (rarely indicated)

MORTON'S NEUROMA (FIGURE 4–139)

General

- Irritation and degeneration of the distal interdigital nerves in the toes from the plantar nerve with eventual enlargement due to perineural fibrosis. This mass can produce pain in the web spaces between the metatarsal heads.
- Most commonly affects the third intermetatarsal space (between the third and fourth digits), followed by the second intermetatarsal space.
- Affects females > males

Clinical Features

- Sharp shooting forefoot pain radiating to the affected digits. Dysesthesias and numbness are common.
- 🕮 Exam: Apply direct pressure to the interdigital web space with one hand and then apply lateral and medial foot compression to squeeze the metatarsal heads together.
- Isolated pain on the plantar aspect of the web space is consistent with Morton's neuroma.

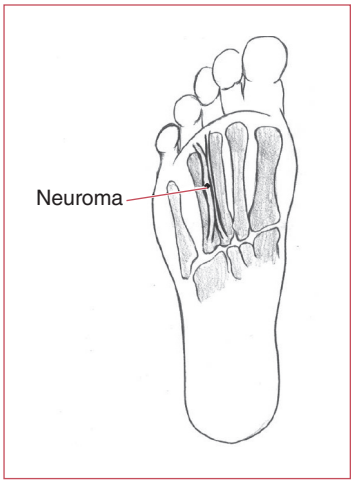

FIGURE 4–139 Morton's neuroma, a perineural fibrosis of the interdigital nerves.

Imaging

- Diagnosis typically based on clinical presentation.
- X-ray to rule out other potential etiology of pain.
- Morton's neuroma can also be confirmed by US and MRI.

Treatment

- *Conservative:*
  - Shoe modifications: Adequate insole cushioning, wide toe box, low heel height
  - Accommodative padding: Metatarsal pads (neuroma pads)
  - Corticosteroid injection may be diagnostic and therapeutic.
- *Surgical:* Excision if indicated

HALLUX DISORDERS: METATARSAL PHALANGEAL SPRAINS, HALLUX VALGUS, AND HALLUX RIGIDUS

General

- Definitions
  - **MTP sprain:**
    - Also known as "turf toe" and is commonly seen in athletes
    - Acute injury to the ligaments and capsule of the first MTP joint
    - Chronic sprains may lead to hallux rigidus.
  - **Hallux valgus:**
    - Lateral deviation of the first toe > normal angle of 15 degrees between the tarsus and metatarsus
    - This may eventually lead to a painful prominence of the medial aspect of the MTP joint (bunion).
  - **Hallux rigidus:**
    - Degenerative joint disease of the first MTP joint leading to pain and stiffness (great toe arthritis of MTP joint)
    - Affects female >> males

Clinical Features

- **MTP sprain:** Acute onset of pain, tenderness, and swelling of the MTP joint, particularly over the plantar aspect. Pain on passive dorsiflexion.
- **Hallux valgus:** Lateral deviation of the first toe with a prominent medial eminence of the MTP joint.
- **Hallux rigidus:** Pain and swelling with decreased ROM of the MTP joint. Antalgic gait pattern.
- **Lesser toe deformities:** The second toe usually will result in an overriding position.

Imaging

- Plain films

Treatment

- *Conservative:* RICE, taping, proper footwear or shoe modifications (e.g., high toe box, forefoot rocker bottom)
- *Surgical:* Debridement (hallux rigidus)

TOE DISORDERS: HAMMER TOE, CLAW TOE, AND MALLET TOE

HAMMER TOE (FIGURE 4–140)

General

- Deformity of the lesser toes in which there is flexion of the PIP joint
- A passive extension of the MTP joint occurs when the toe is flat on the ground. The DIP joint is usually not affected.
- Caused by chronic, tight shoe wear that crowds the toes, but may be seen after trauma.

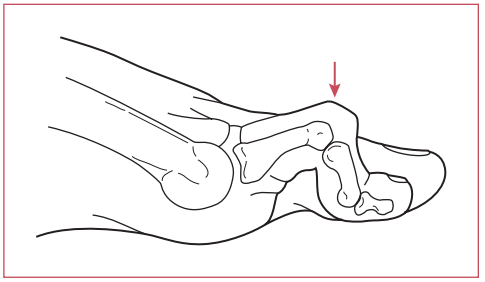

FIGURE 4–140 Hammer toe.

Clinical Features

- Obvious deformity as described
- Pain is present in the toe and the patient has difficulty with footwear.

Imaging

- Standing AP and lateral x-ray films help exclude other diagnoses.

Treatment

- Shoes with high toe boxes. Shoes should be 1/2 inch longer than the longest toe. Custom foot orthotics.
- Home exercise program of passive manual stretching

CLAW TOE (FIGURE 4–141)

General

- Characterized by extension of MTP, flexion of the PIP, and flexion of the DIP
- Deformity is usually the result of the incompetence of the foot intrinsic muscles, secondary to the neurologic disorders affecting the strength of these muscles (i.e., diabetes, alcoholism, peripheral neuropathies, Charcot–Marie–Tooth disease, and spinal cord tumors).

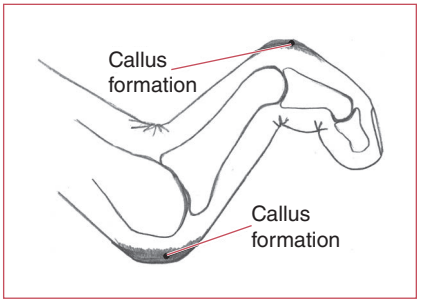

FIGURE 4–141 Claw toe.

Clinical Features

- Pain is the principal symptom.
- If progression is rapid, this suggests a related neurologic condition.

Imaging

- Radiographs of foot to confirm the diagnosis

Treatment

- Shoes with soft insoles and high toe boxes
- Splints available
- Surgical correction may be necessary if conservative treatment fails.

MALLET TOE (FIGURE 4–142)

General

- Flexion deformity at the DIP joint with normal alignment at the PIP and MTP joints
- Usually the result of jamming type injury or wearing tight shoes

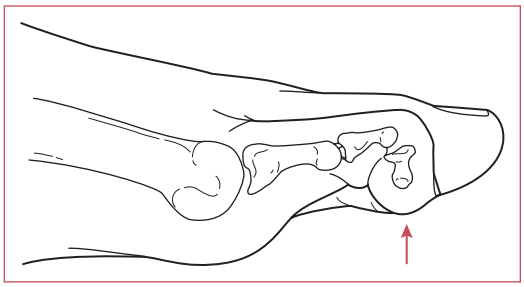

FIGURE 4–142 Mallet toe.

Clinical Features

- Obvious deformity, pain, callus at tip of toe

Imaging

- AP and lateral x-rays may be indicated to rule out associated avulsion fracture.

Treatment

- The callus should be trimmed.
- Shoes with soft insoles and high toe boxes are used.
- Surgical treatment includes flexor tenotomy. If the deformity is fixed, condylectomy is required.

LISFRANC JOINT INJURY

General

- Spectrum of midfoot injuries, from sprains to fracture/dislocations at the tarsometatarsal joint, which is also known as the Lisfranc joint **(Figure 4–143)**

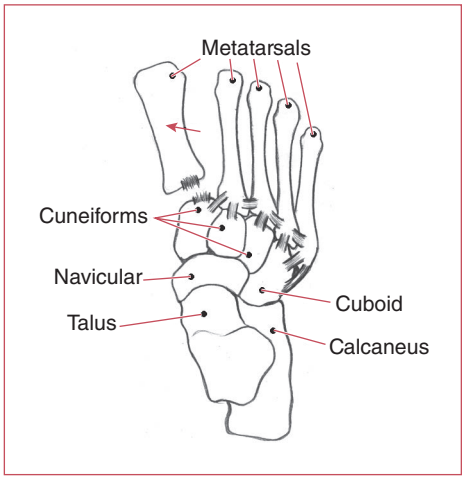

FIGURE 4–143 Isolated Lisfranc dislocation.

Mechanism of Injury

- Low-energy trauma: Caused by a direct impact to the joint or by axial loading of the midfoot and rotating it
  - Commonly seen in athletes
- High-energy trauma: Less common. Due to direct, high-impact trauma (e.g., MVA) with greater damage produced.

Clinical Features

- Vague foot or ankle pain. Pain and swelling localized to the dorsum of the foot.
- This injury is easily missed and often misdiagnosed as a lateral ankle sprain.
- Pain may be exacerbated by stabilizing the hindfoot and rotating the forefoot.

Imaging

- X-rays: AP, lateral, and oblique views of the ankle and the entire foot. Look for a shift commonly between the first and second metatarsals.
- MRI or CT if needed

Treatment

- *Conservative:*
  - Nondisplaced joint: Non-weight bearing, immobilization for 6 to 8 weeks with continued support thereafter
- *Surgical:*
  - Stabilization is integral to maintaining the bony architecture of the entire foot.

FOOT FRACTURES

General

- The fifth metatarsal is the most common metatarsal fractured.
- Fractures at the base of the fifth metatarsal can be classified by zone:
  - **Zone 1:** Pseudo-Jones fracture—avulsion fracture of the tuberosity
  - **Zone 2:** Jones' fracture occurs at the metaphyseal–diaphyseal junction. Risk of nonunion.
  - **Zone 3:** Diaphyseal stress fractures typically result from repetitive loading. Risk of nonunion.
- **Dancer's fracture:** Distal shaft fracture of the fifth metatarsal
- **Nutcracker fracture:** Cuboid fracture
- **March fracture:** Metatarsal stress fracture

Clinical Features

- Pain with palpation; swelling and ecchymosis over the involved area
- Usually a result of trauma or repetitive stress

Imaging

- Plain films of foot and ankle; MRI or CT

Treatment

- Jones' fracture:
  - Non-weight bearing cast for 6 weeks
  - ORIF if nonunion occurs
- Nutcracker fracture: ORIF
- March fracture:
  - Relative rest with immobilization
  - Cast if needed
  - March fractures of the fifth metatarsal may require surgical fixation due to the increased risk of fracture displacement.

TURF TOE

General

- Sprain of the first MTP joint capsule by forced hyperextension
- Commonly occurs when athletes play on unyielding artificial surfaces with flexible shoes

Clinical Features

- Pain is reproduced by passive extension of the first MTP with pain at the joint capsule.

Imaging

- AP and lateral radiographs may be indicated to rule out fracture with attention to the sesamoid.

Treatment

- Firmer toe box shoes, taping, immobilization by first metatarsal splints, and/or use of orthoplast inserts

JOINT INJECTIONS AND ASPIRATIONS

Indications for Aspiration

- **Diagnostic:**
  - Joint fluid analysis
  - Rule out infection

  - Determine the difference between inflammatory and OA
  - Evaluate for crystalline arthropathies
  - Presence of blood may indicate trauma (i.e., ACL tears, osteochondral fractures)
- **Therapeutic:**
  - Fluid aspiration temporarily decreases pain.

Indications for Injection

- **Diagnostic:**
  - Anesthetics may reduce pain from specific structures.
- **Therapeutic:**
  - Corticosteroids may reduce inflammation in specific structures.

Contraindications for Needle Aspirations and Injections

GENERAL CONTRAINDICATIONS

- Infection:
  - Local: Cellulitis of the overlying skin
  - Systemic: Bacteremia or sepsis
- Coagulopathy:
  - As caused by anticoagulation medications
  - Prolonged bleeding times
  - Genetic coagulopathies
- Lymphedema—in severe cases of edema at the site of injection
- Skin disorders:
  - Overlying psoriatic plaques at the site
- Contraindications **for use of anesthetic (i.e., lidocaine):**
  - Allergy to lidocaine family of anesthetics
- Contraindications **for use of corticosteroids:**
  - Corticosteroid allergy
  - Infection: Cellulitis, septic arthritis, or osteomyelitis of adjacent bone
  - Immunocompromised patient
  - Acute monoarticular arthritis of unknown etiology
  - Directly prior to a surgical implant
  - Postsurgical joint implant
  - Osteochondral fracture (impedes healing)
  - Severe osteoporosis in adjacent bones
  - Charcot joint (increases risk of AVN)

Possible Side Effects of Corticosteroids

- **Local:**
  - Infection
  - Subcutaneous fat atrophy
  - Skin depigmentation
  - Tendon rupture
- **Systemic:**
  - Skin flushing
  - Menstrual irregularity (high doses)
  - Impaired glucose tolerance
  - Osteoporosis (prolonged use)
  - Psychosis (high doses)
  - Steroid arthropathy
  - Adrenal suppression
  - Immunosuppression (high, chronic doses)
  - Avascular necrosis

- **Anesthetic side effects:**
  - Overdose symptoms:
    - Early signs: Tingling around lips and tongue
    - Later signs: Convulsions, coma, respiratory arrest
  - Allergic symptoms:
    - Mild: Flushing, itching, urticarial
    - Advanced: Chest tightness, abdominal pain, nausea, vomiting
    - Catastrophic: Anaphylaxis, circulatory collapse, death

Corticosteroids

- Corticosteroids vary in strength, duration, and side effects.
- Some commonly used forms include methylprednisolone, triamcinolone, and betamethasone.
- 🕮 Triamcinolone is more likely to cause tissue atrophy than methylprednisolone when injecting superficial structures.

🕮 Viscosupplementation/Hyaluronic Acid Injections

- Hyaluronan or hyaluronic acid (HA) is a large, linear glycosaminoglycan and is a major component of the synovial and cartilage extracellular matrix. It is important for joint lubrication, tissue hydration, and protein homeostasis.
- Benefits believed to be derived from enhanced endogenous HA synthesis by synovial cells, proteoglycan synthesis by chondrocytes, anti-inflammatory effects, and analgesic effects
- Several formulations of hyaluronan and high molecular weight hyaluronan have been Food and Drug Administration (FDA)-approved to treat symptomatic OA of the knees that has failed more conservative measures. These formulations range from one injection to a series of 3 to 5 weekly injections.
- Use caution with patients with allergy to products from birds such as feathers, eggs, or poultry.
- Side effects are the same as with any type of injection.
- Duration of pain relief can last as long as 6 months.

Platelet Rich Plasma Injections

- Platelet rich plasma (PRP) refers to plasma that is concentrated with platelets and growth factors to help stimulate a repair response in various musculoskeletal injuries. It can be used to help the healing process in tendons, ligaments, and joints by recruiting stem cells and increasing vascularity to the injured region.
- PRP is derived from the patient's blood by using a centrifuge to separate out blood products into layers, and then red blood cells and platelet poor plasma are removed. It is injected into the tendon or ligament structure under image guidance.

COMMON INJECTION TECHNIQUES

Shoulder Injections

Acromioclavicular Joint Injection

- Identify the superior tip of the acromion—the joint line is approximately 2 cm medial to the acromion.
- Insert the needle from the superior position, angling about 30 degrees medially.

Subacromial Bursa Injection

- Lateral approach:
  - Locate the lateral edge of the acromion.
  - Insert the needle at the midportion of the acromion, angling slightly upward.
- Posterolateral approach:
  - Locate the posterolateral aspect of the acromion.
  - Insert the needle at the midportion of the acromion, angling slightly upward under the acromion.

Elbow

Lateral Epicondyle Injection

- Support the elbow bent at 90 degrees and the forearm supinated.
- Palpate the origin of the ECR-B distal to the lateral epicondyle.
- Identify the facet lying anteriorly on the lateral epicondyle.
- Insert the needle in line with the cubital crease perpendicular to the facet until it touches the bone and slightly retract.
- Beware the radial nerve.

Medial Epicondyle Injection

- Support the arm extended.
- Identify the anterior facet on the medial epicondyle.
- Insert the needle perpendicular to the facet until it touches bone.
- Beware the ulnar nerve.

Olecranon Bursa Injection

- Identify the bursa at the tip of the olecranon.
- In aspirating the olecranon bursa, some practitioners use a zigzag motion on needle insertion to avoid developing a sinus tract.

Hand

First Carpometacarpal Joint Injection

- Rest the hand at the mid-position with the thumb up.
- Identify the joint space between the metacarpal and the trapezium at the apex of the anatomic snuff box.
- Insert the needle at a 90-degree angle to the surface at the joint space.

De Quervain's Tenosynovitis Injection

- Place the hand vertical with the thumb in slight flexion.
- Identify the APL and EPB tendons. ("**AP**p**LE** & **P**eanut **B**utter")
- Slide the needle into the gap between the two tendons.

Trigger Finger Injection

- Identify the tendon nodule.
- If the needle moves with finger flexion, withdraw slightly until the needle is still inside the tendon sheath but external to the tendon.

Hip

Greater Trochanteric Bursa Injection

- Position the patient on the side with the affected trochanter up, the upper leg extended, and the lower leg flexed.
- Insert the needle at the center of the tender area to the bone and withdraw 1 to 2 mm.

Knee

Knee Joint Injection

- The medial approach under the patella may provide more space for the needle than the lateral approach.
- Insert the needle between the midpoint of the medial edge of the patella and the femoral condyle, sliding the needle under the patella.

- The superolateral patellar (lateral suprapatellar) approach is another method of injection into the knee when performed blindly or under US guidance.
- With the patient supine and the knee extended, identify the patellar tendon. Insert the needle approximately one fingerbreadth superior and one fingerbreadth lateral to the patella and slide the needle underneath the patella into the suprapatellar bursa.
- Medial patellofemoral and arthroscopic approaches (anterolateral and anteromedial), which utilize the same entry points used for knee arthroscopy, are also alternative approaches.

Pes Anserine Bursa Injection

- Identify the bursa by having the patient flex the knee against resistance and palpating the medial hamstring tendons distally at the insertion on the tibia.
- The bursa is found as an area of extreme tenderness underneath the tendons near their attachment to the tibia.
- Insert the needle into the center of the area to the bone and withdraw 1 to 2 mm.

Ankle

Tibiotalar Joint Injection

- With the foot in neutral, passively flex and extend the ankle, palpating for the joint at the small triangular space at the lateral side of the ankle.
- Insert the needle into the joint, directing it toward the midpoint of the tibia.

Foot

Morton's Neuroma Injection

- The most common injection location is between the third and fourth metatarsal heads.
- Place the needle into the dorsal foot in line with the MTP, 1 to 2 cm proximal to the web space.
- Advance the needle into the plantar aspect of the foot until the skin slightly tents, then withdraw approximately 1 cm into the neuroma.

Plantar Fasciitis Injection

- Identify the tender area on the heel, usually distal to the medial aspect of the calcaneus.
- Insert the needle into the medial side of the soft part of the sole of the foot above the fat pad.
- Advance the needle to the calcaneal–fascial juncture.
- Beware of complications: Footpad atrophy and plantar fascia rupture.

TRIGGER POINTS

Definition

- A hyperirritable point within a taut band of skeletal muscle or fascia that is sensitive and can be painful to digital compression with a consistent reproducible referred pain pattern.
- Active trigger points can actively refer pain at rest.
- Latent trigger points do not actively refer pain at rest.
- Referred pain patterns may be elicited with direct pressure.
- May restrict motion and cause weakness

Location

- Can be myofascial, cutaneous, fascial, ligamentous, or periosteal in origin
- Pain radiates from a trigger point into a specific reproducible zone of reference.

Causes

- Acute trauma or repeated microtrauma
- Muscle stress and imbalance from prolonged static postures
- Lack of dynamic exercise
- Chronic muscle contraction may be caused by uncontrolled acetylcholine release.

Treatment

- Treatment should be directed toward a comprehensive rehabilitation program for optimal results:
  - Physical therapeutics, modalities (heat/ice, US, transcutaneous electrical nerve stimulation [TENS]), spray (freeze), and stretch, massage, postural alignment, and acupressure (ischemic compression therapy)
- Trigger point injections may be used as an adjunct to this treatment regimen.
- A number of trigger point injection methods for myofascial trigger points have been tried with varying results, including the following:
  - Dry needle insertion with peppering of the tender zone
  - Injection of local anesthetic alone
  - Injection of local anesthetic mixed with corticosteroids
  - Inconclusive evidence to support the use of botulinum toxin in treating myofascial pain
  - Side effects of injection of corticosteroids and botulinum toxin may include myositis.

SPINE REHABILITATION (ALSO SEE PAIN OF SPINAL ORIGIN AND INTERVENTIONAL SPINAL PROCEDURES SECTIONS IN CHAPTER 11, PAIN MEDICINE)

🕮 INTRODUCTION

- Neck and back pain are the leading musculoskeletal disorders that contribute to impairment and disability:
  - Injury to the lumbar region in particular has up to a 15% (Deyo et al., 2002; Hoy et al., 2010) annual incidence and 84% (Walker, 2000; Manchikanti, 2002; Dagenais et al., 2008; Thiese et al., 2014) lifetime prevalence (70% cervical, 15% thoracic), affecting more than 100 million people in the United States alone.
- Fortunately, the natural course is favorable, as symptoms are usually self-limited. Though resolution is likely, recurrence of symptoms is possible because of structural and pathologic functional adaptations. These can be addressed and limited with adequate comprehensive treatment programs.
- This section focuses on board-related topics with regard to spinal disorders and is to be used as a study guide. It is not intended to be an all-inclusive composite. For more elaborate coverage of the subject matter, the reader is directed to the suggested references at the end of this chapter.

🕮 Clinical Course

- Disability and the ability to return to work generally improve in 1 month, but one-third may have persistent discomfort for up to a year after injury, with 20% of those reporting limitation in activity.

OUTCOME	TIME
Approximately 50% resolve	Approximately 1–2 weeks
Approximately 90% resolve	Approximately 6–12 weeks
Approximately 85% recur	Approximately 1–2 years

- Approximately 10% of patients with low back pain continue with residual complaints. Due to its morbidity, this subgroup constitutes the second most common reason for primary care physician office visits.
- Proper treatment of these patients depends on an accurate diagnosis, which may be elusive due to the complexity of the structures involved.

- Diagnostic testing may be indicated to further define pathology and to focus care.
- Regardless, proper screening begins with a complete history and physical examination, assessment for the presence of red flags (conditions requiring more immediate attention), and a comprehensive treatment plan.

Red Flags

CLINICAL PRESENTATION	CONDITION
Gait ataxia/upper motor neuron changes	Myelopathy
Bowel/bladder/sexual dysfunction	Cauda equina syndrome/myelopathy
Night pain/weight loss	Tumor
Fevers/chills	Infection

- In the workforce, low back pain is second only to upper respiratory infections as the most frequent cause of work absenteeism.
- Due to the cost of medical care, time lost from work, disability payments, production loss, staff retraining, and litigation expenses, its economic impact reaches into the billions annually:
  - Approximately 25 million Americans lose one or more days from work a year due to back pain.
  - Over five million people are disabled from low back pain and the yearly prevalence continues to grow at a rate greater than that of the size of the general U.S. population.
- Those who develop chronic low back pain cause 80% to 90% of healthcare expenditures.

Absenteeism

TIME MISSED FROM WORK	RETURN TO WORK EXPECTATIONS (%)
APPROXIMATELY 6 MONTHS	50
APPROXIMATELY 1 YEAR	25
APPROXIMATELY 2 YEARS	0

FUNCTIONAL ANATOMY

Cervical Vertebrae

Atypical: C1 and C2

UNIQUE CHARACTERISTICS

- **C1 Vertebra (Atlas; Figure 4–144):**
  - Ring-shaped bone containing two lateral masses
  - No vertebral body or spinous process

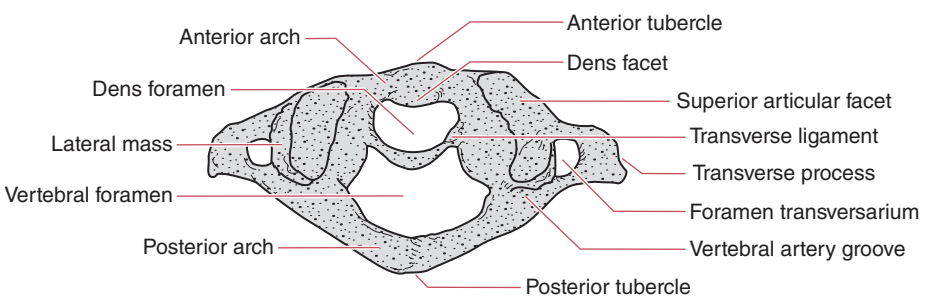

FIGURE 4–144 The atlas, superior view.

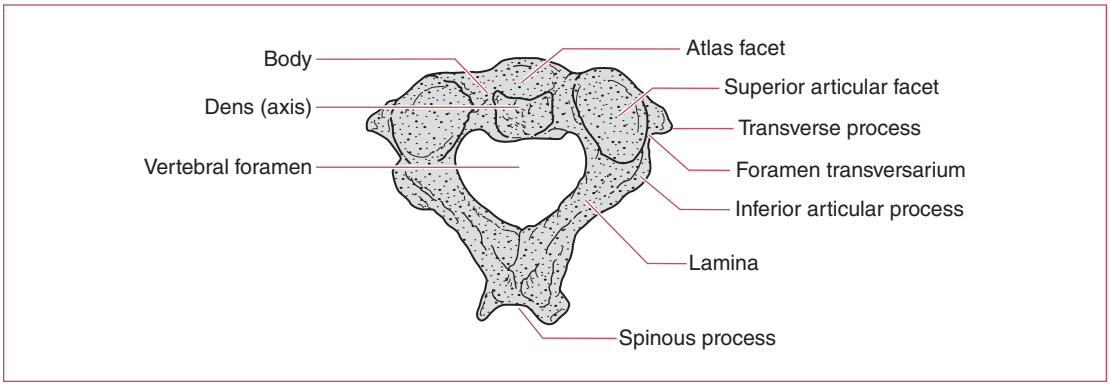

FIGURE 4–145 The axis, superior view.

- **C2 Vertebra (Axis; Figure 4–145):**
  - Its vertebral body has an odontoid process.
  - Bifid spinous process (C2–C6 vertebrae have bifid spinous processes)

Typical: C3–C7 Vertebrae (Figure 4–146)

UNIQUE CHARACTERISTICS

- **Anterior region:**
  - 🕮 Presence of vertebral bodies
  - Cervical uncinate processes: Raised spondylotic margins along the lateral aspect of the superior surface of a cervical vertebral body due to disc degeneration. These raised margins approximate with the body of the superior vertebra, creating a degenerative joint known as an uncovertebral joint (joint of Luschka).
  - The joints of Luschka function to limit lateral translation (Figure 4–147).

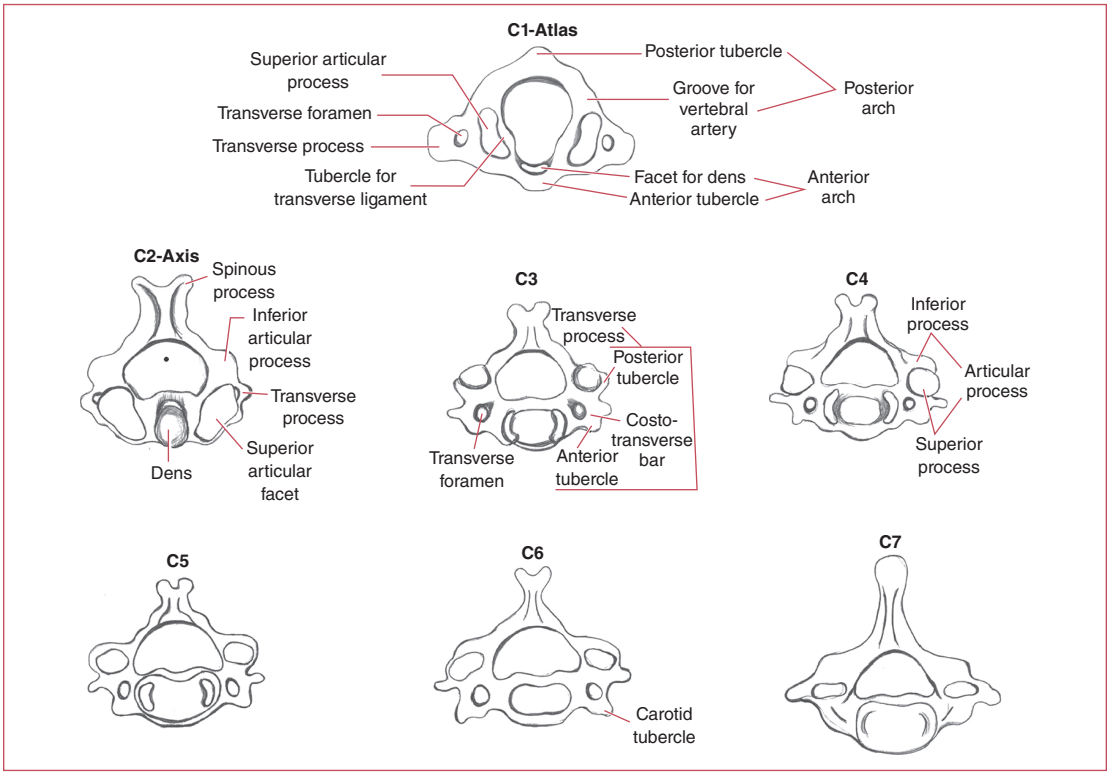

FIGURE 4–146 Typical cervical vertebrae—superior view.

- **Posterior region:**
  - Pedicles, superior articular processes (SAPs), inferior articular processes (IAPs), laminae, transverse processes (TPs), foramen transversarium, and spinous processes:
    - C3, C4, C5, C6: Bifid spinous processes
    - C7: Nonbifid spinous process

Thoracic Vertebrae

T1–T12 (Figure 4–148)

UNIQUE CHARACTERISTICS

- **Anterior region:**
  - Vertebral bodies with articulations for the rib heads
- **Posterior region:**
  - Pedicles, SAP, IAP, laminae, TPs with articulations for rib tubercles and spinous processes

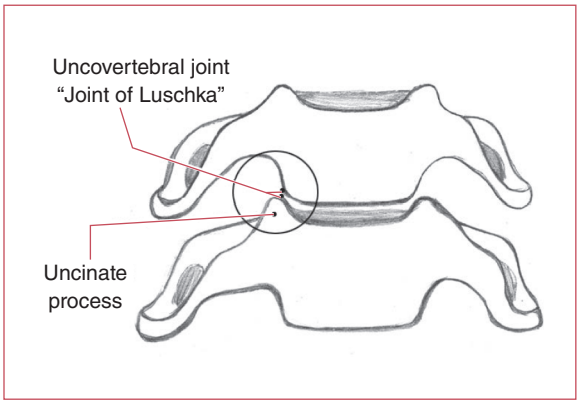

FIGURE 4–147 Joints of Luschka.

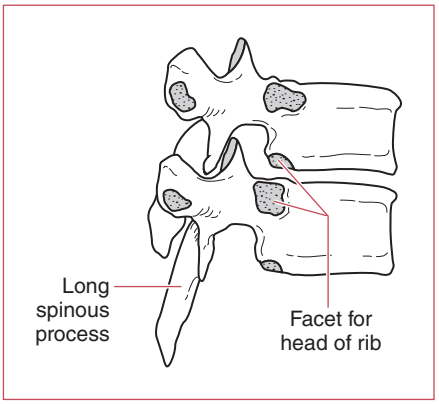

FIGURE 4–148 Thoracic vertebrae—lateral view.

Lumbar Vertebrae

L1–L5 (Figure 4–149)

UNIQUE CHARACTERISTICS

- **Anterior region:**
  - Presence of vertebral bodies
- **Posterior region:**
  - Pedicles, SAP, IAP, TPs, mammillary processes, laminae, and spinous processes

Lumbosacral Transitional Vertebrae

- Congenital anatomic variant in the lumbosacral spine that occurs with a prevalence of 4% to 30% in the general population (Konin & Walz, 2010).
- The importance of identifying lumbosacral transitional vertebrae (LSTV) in patients is for correctly identifying the level of pathology, notably when planning for an interventional spinal procedure or surgery.
- Nomenclature:
  - **Sacralization** is an anomalous partial or complete fusion of the L5 vertebra to the sacrum. Incidence: Approximately 1% complete, approximately 6% incomplete
  - **Lumbarization** refers to an anomalous partial or complete nonunion of the S1 segment of the sacrum. This forms an additional lumbar segment ("L6" vertebra) and leaves four remaining fused sacral segments. Incidence approximately 4%.

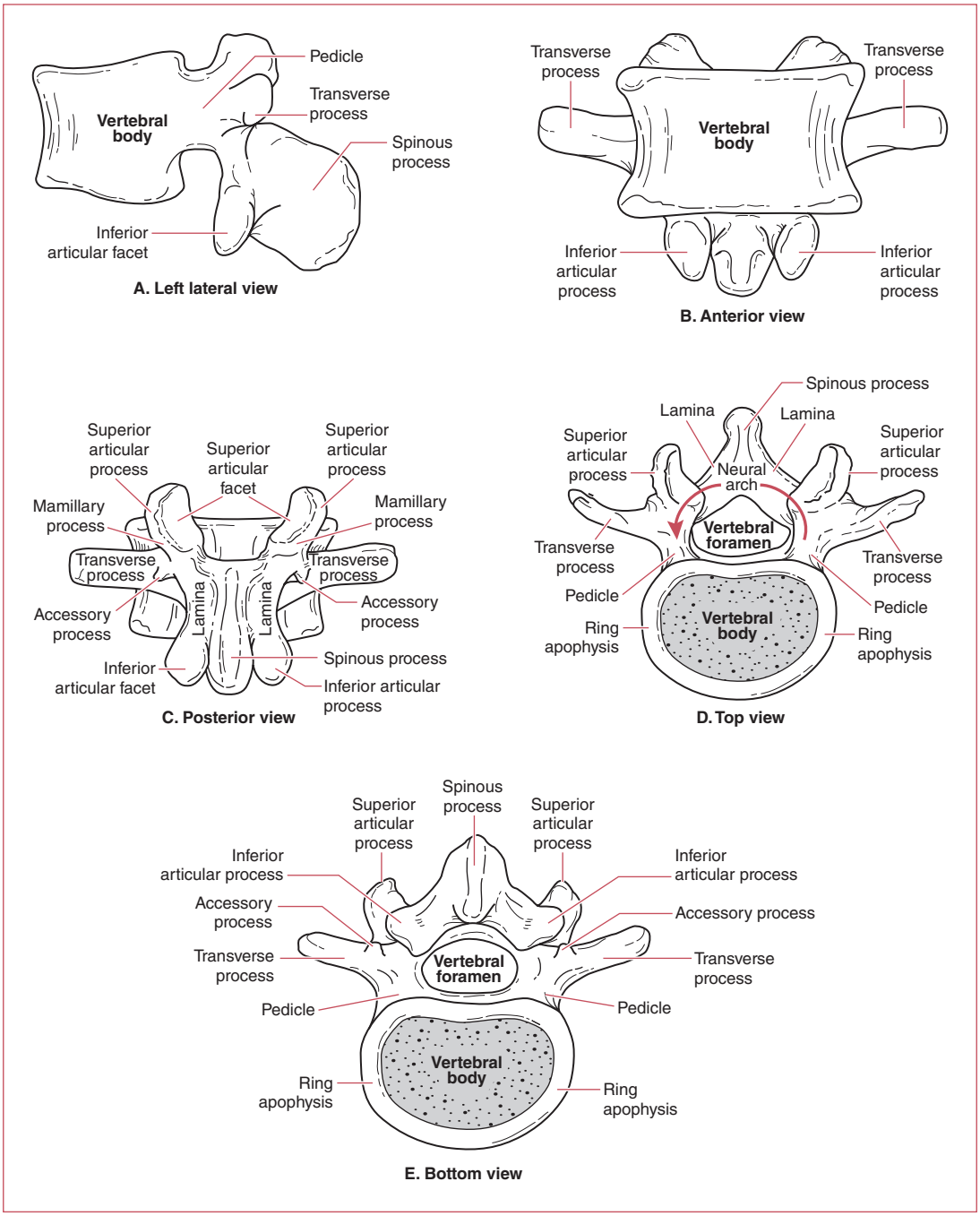

FIGURE 4–149 Lumbar vertebrae—five views. (A) Left lateral view. (B) Anterior view. (C) Posterior view. (D) Top view. (E) Bottom view.

Spinal Motion Segment: The Three-Joint Complex (Figure 4–150)

- A three-joint complex is formed between two lumbar vertebrae to create a motion segment.

Joint 1	Vertebral body endplate–disc–endplate joint
Joint 2	Zygapophyseal joint (facet joint)
Joint 3	Zygapophyseal joint (facet joint)

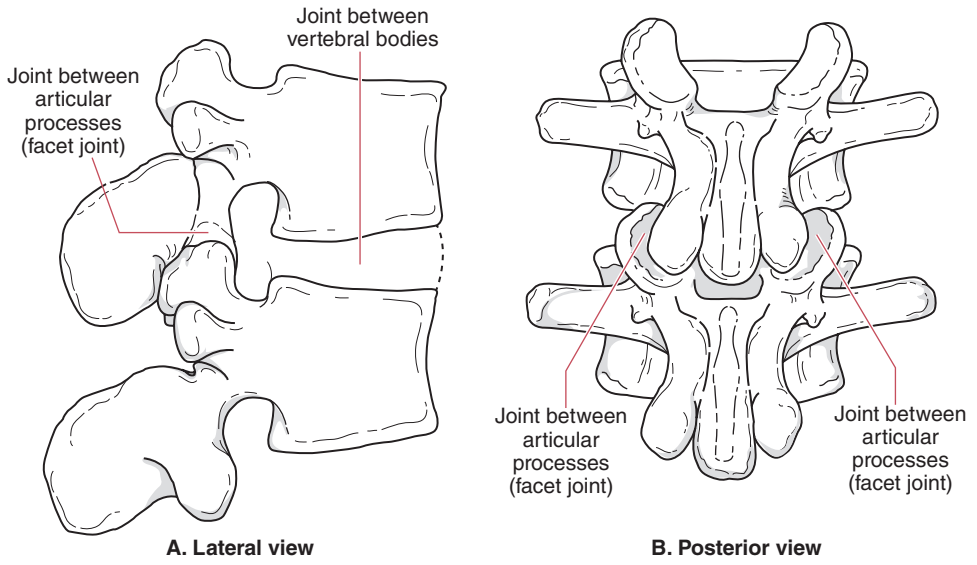

FIGURE 4–150 The three-joint complex. (A) Lateral view of lumbar vertebrae. (B) Posterior view of lumbar vertebrae.

Sacral Vertebrae

🕮 S1–S5 (Figure 4–151)

UNIQUE CHARACTERISTICS

- A triangular-shaped bone consisting of five fused vertebrae (S1–S5)
- Four pairs of foraminae (anterior and posterior), sacral promontory, sacral ala, hiatus, cornua, medial, intermediate, and lateral crests, which are analogous to the spinous processes
- Sacral ligaments: See **Figure 4–152**.

Coccygeal Vertebrae

- Three to four fused segments, with TPs, hiatus, and cornua

Facet Joints

Characteristics (Figure 4–153)

- Also known as zygapophyseal (ZP- or Z-) joints, apophyseal joints
- The facet joint is a true synovial joint composed of:
  - SAP
  - IAP
  - Joint capsule
  - Articular cartilage
  - Meniscus

🕮 Facet Joint Orientation

- Cervical:
  - Atlanto-axial (AA) and atlanto-occipital (AO) joints have no true facet joints due to their atypical anatomy.
  - C3–C7 facets are positioned in the frontal (coronal) plane.
- Thoracic facets are also positioned in the frontal (coronal) plane.
- Lumbar facets begin in the sagittal plane in the upper lumbar region and progress to the frontal plane at L5–S1.

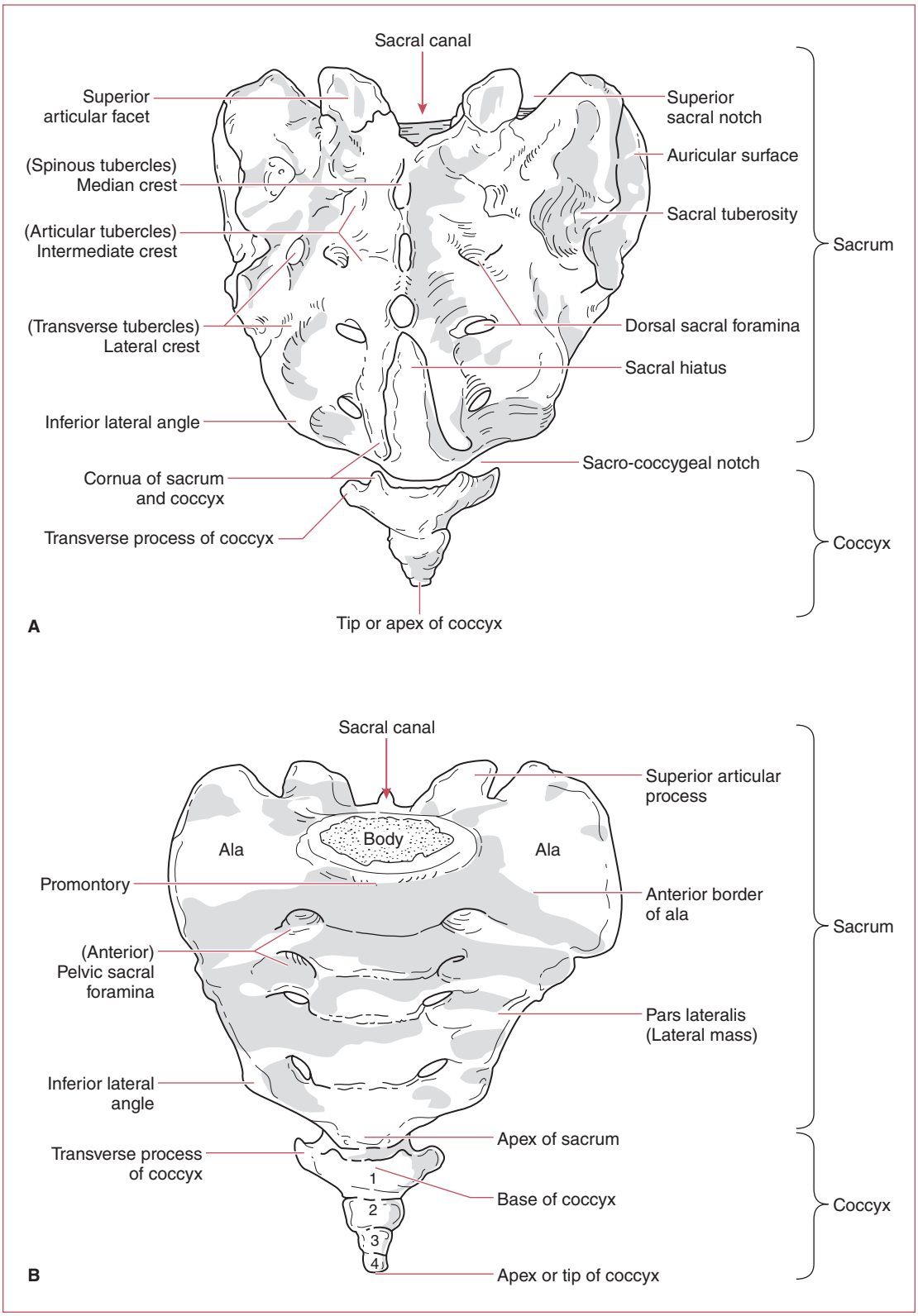

FIGURE 4–151 The sacrum and coccyx. (A) Dorsal surface. (B) Pelvic surface.

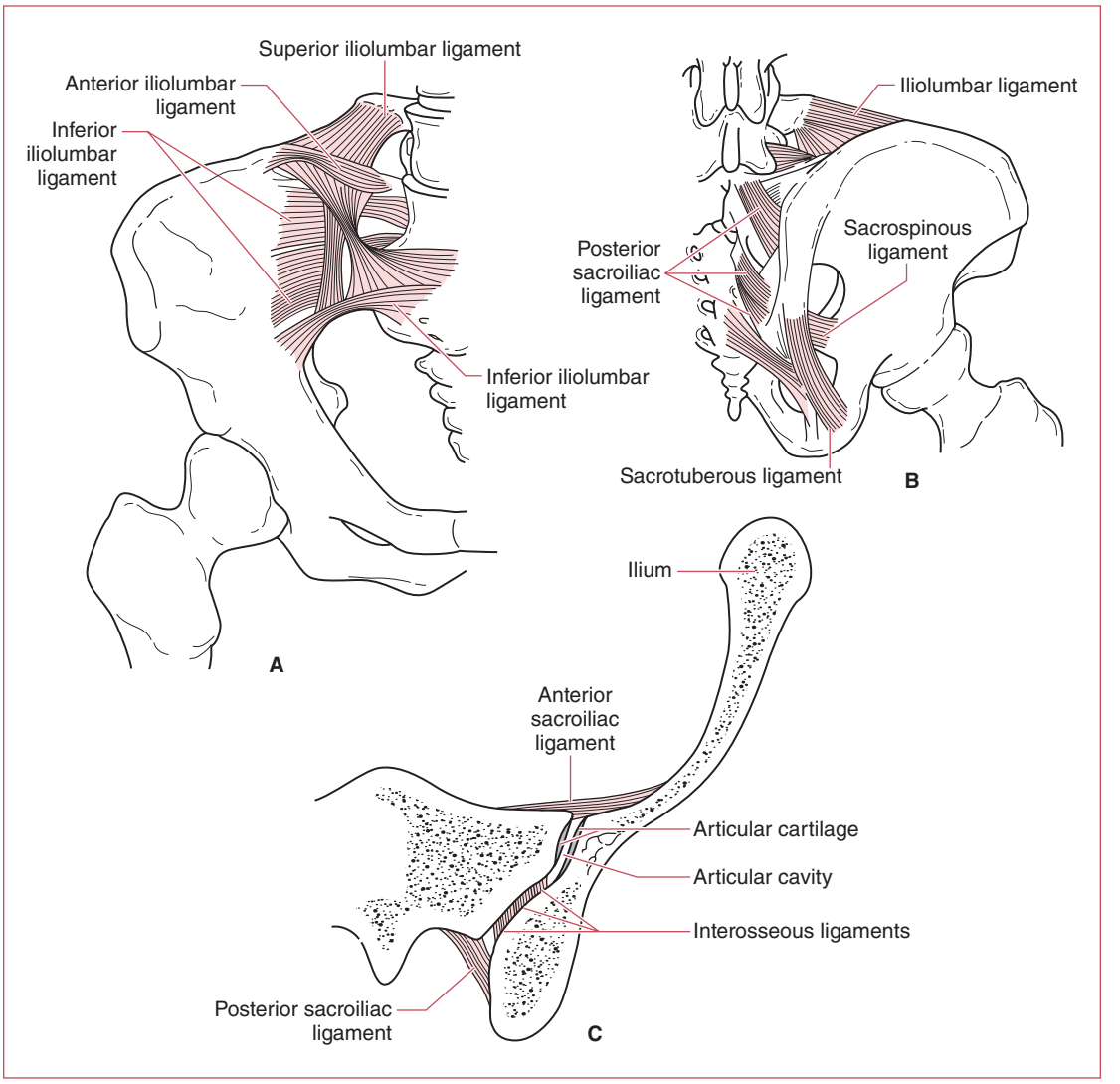

FIGURE 4–152 Lumbosacral ligaments. (A) Anterior view. (B) Posterior view. (C) Transverse view.

Facet Innervation

- Each facet joint is innervated by two medial branch nerves from spinal nerve dorsal rami. The pattern of innervation differs depending on the spinal region.
- Facet joints in the cervical spine are innervated by medial branches from spinal levels of that facet:
  - Example: C5–C6 facet is innervated by C5 and C6 medial branches.
- However, facets in the thoracic and lumbar spines are innervated by medial branches from the top spinal level and the level above:
  - Example: L4–L5 facet is innervated by the L3 and L4 medial branches.

Function

- Limit vertebral motion
- Resist shearing and rotational forces
- Weight bearing: Increases with spinal extension and with decreased disc heights

🕮 THE INTERVERTEBRAL DISC

Characteristics (Figure 4–154)

- **Nucleus pulposus:** A viscous mucoprotein gel mixture of water and proteoglycans in a network of Type II collagen that braces the annulus to prevent buckling.
- **Annulus fibrosus:** Type I collagen fibers arranged in obliquely running lamellae that encase the nucleus pulposus and are attached to the vertebral endplate plates. This orientation withstands distraction forces and bending but is more susceptible to injury with torsional stresses.
- **Vertebral endplate:** Cartilaginous covering of the vertebral body apophysis, forming the interface between the disc and vertebral body (forming the top and the bottom of the disc).

Vascular Supply

- Supplied by cartilaginous vertebral body endplates
- Intervertebral discs are essentially avascular by adulthood.

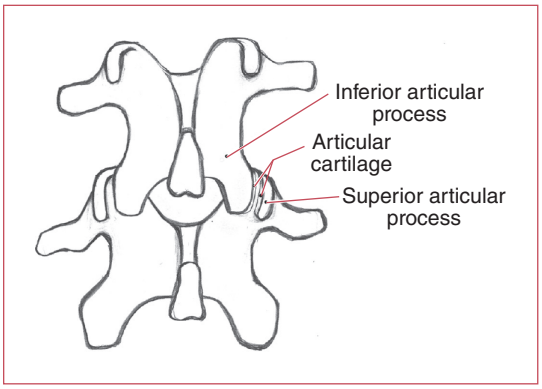

FIGURE 4–153 Lumbar facet joint—posterior view. The posterior capsule has been resected to reveal the joint cavity.

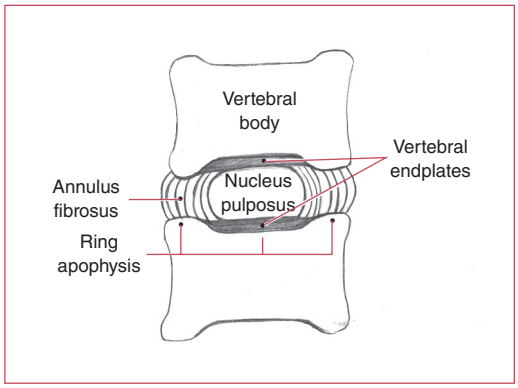

FIGURE 4–154 The intervertebral disc.

Innervation (Figure 4–155)

- The outer one-third of the intervertebral disc contains the annulus fibrosis, which receives innervations from the sinuvertebral nerve and gray ramus communicans, both from the bilateral ventral rami. The nucleus pulposus lacks any innervation.
- The anterolateral part of the annulus fibrosis is innervated by ventral rami and gray rami communicans.
- The posterior part of the annulus fibrosis is innervated by sinuvertebral nerves (recurrent branches off of the ventral rami).

🕮 Innervations (Figure 4–155)

NERVE	CONTRIBUTIONS
Ventral primary rami	Trunk musculature, plexus contributions
Dorsal primary rami	Lateral: Iliocostalis, skin Intermediate: Longissimus Medial: Multifidi, rotators, interspinalis, intertransversarii, posterior spinal ligaments, zygapophyseal joints
Sinuvertebral nerve	Posterior longitudinal ligament (PLL), posterior disc, anterior dura, vertebral body

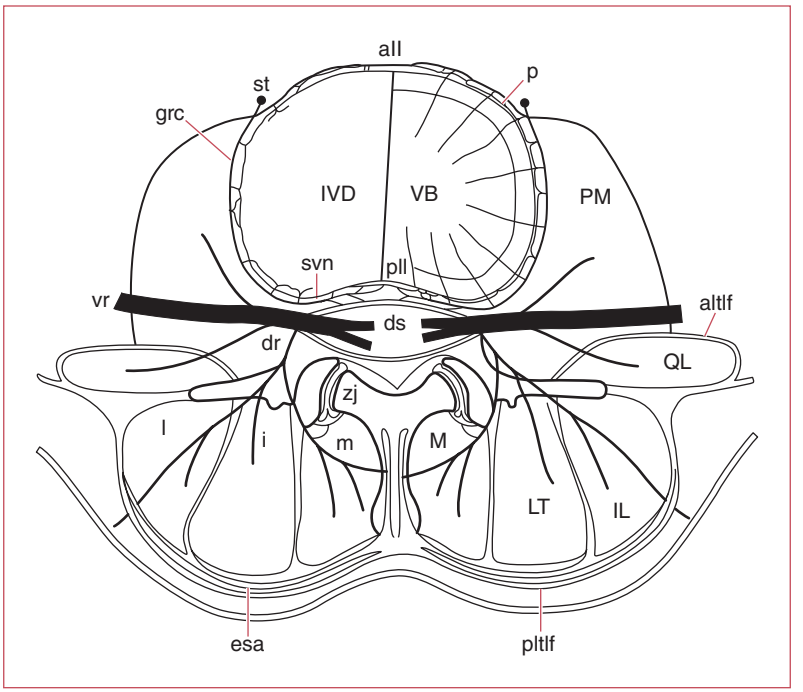

FIGURE 4–155 The lumbar spine innervations. Cross-sectional view, which incorporates the level of the VB and the P on the right and the IVD on the left.
all, anterior longitudinal ligament; altlf, anterior layer of thoracolumbar fascia; dr, dorsal ramus; ds, dural sac; esa, erector spinal aponeurosis; grc, gray ramus communicans; i, intermediate branch; IL, iliocostal lumborum; IVD, intervertebral disc; l, lateral branch; LT, longissimus thoracis; m, medial branch; M, multifidus; p, periosteum; pll, posterior longitudinal ligament; pltlf, posterior layer of thoracolumbar fascia; PM, psoas muscle; QL, quadratus lumborum; st, sympathetic trunk; svn, sinuvertebral nerve; VB, vertebral body; vr, ventral ramus; zj, zygapophyseal joint.

Function

- Allows for vertebral body motion
- Weight bearing (Figure 4–156)

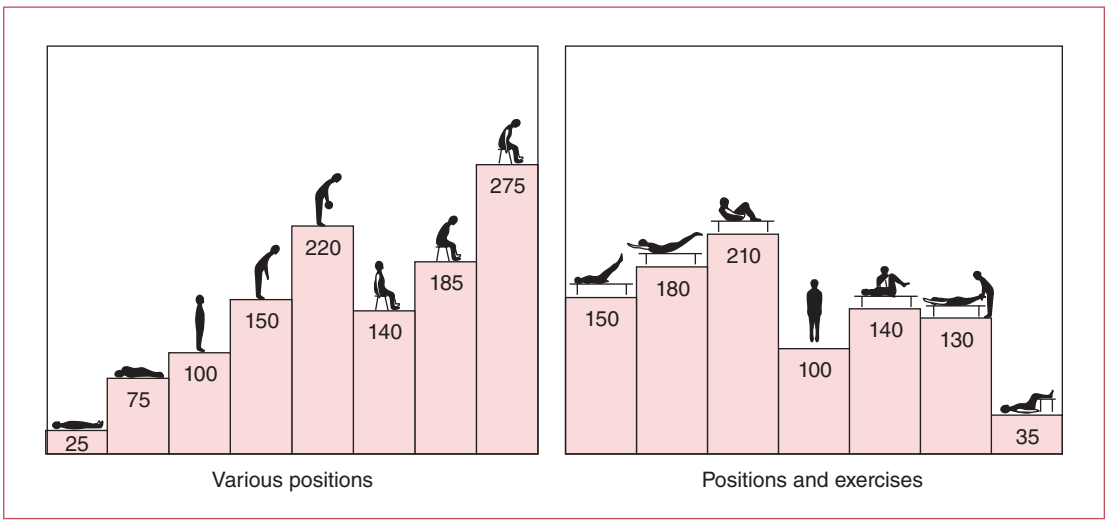

FIGURE 4–156 Positional disc pressure changes. Relative change in pressure (or load) in the third lumbar disc in various positions in living subjects.
Note: Neutral erect posture is considered 100% in the figures; other postures and activities are calculated in relationship to this.
Source: From Nachemson AL. The lumbar spine: an orthopaedic challenge. *Spine*. 1976;1:59–71. doi:10.1097/00007632-197603000-00009, reprinted with permission.

Aging Effects

DECREASES	INCREASES
Nuclear water content	Fibrous tissue
Ratio of chondroitin:keratin	Cartilage cells
Proteoglycan molecular weight	Amorphous tissue

Spinal Ligaments (Figure 4–157)

- **Anterior longitudinal ligament (ALL):**
  - Runs the entire length of the anterior spine, covering the anterior aspect of each vertebral body and disc
  - Function: Limits hyperextension and forward movement
- **Posterior longitudinal ligament (PLL):**
  - Attaches to the posterior rim of the vertebral bodies and disc from C2 to the sacrum. It continues superiorly with the tectorial membrane to the occiput.
  - Function: Prevents hyperflexion of the vertebral column
- **Posterior spinal element ligaments**
  1. Ligamentum flavum
  2. Interspinous ligaments
  3. Supraspinous ligaments

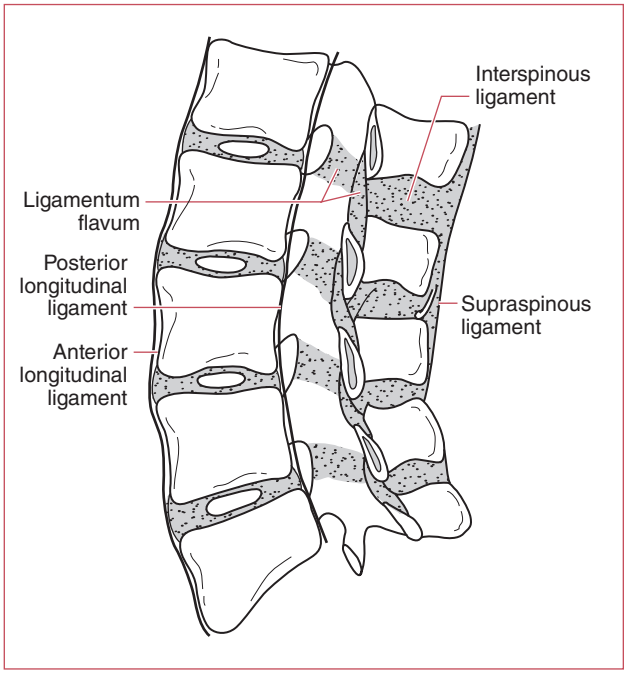

FIGURE 4–157 Lumbar spine ligaments, median sagittal view.

- **Ligamentum flavum:**
  - Elastic ligament that connects adjacent vertebral arches longitudinally, attaching laminae to laminae
  - Function: Maintains constant disc tension and assists in straightening the column after flexion
- **Interspinous and supraspinous ligaments:**
  - Run from spinous process to spinous process. The supraspinous ligament runs from C7 to L3.
  - Function: Weakly resist both spinal separation and flexion
- **Ligamentum nuchae:**
  - Superior continuation of the supraspinous ligament extending from the occipital protuberance to C7
  - Function: Boundary of the deep muscle in the cervical region
- **Intertransverse ligaments (ILs):**
  - Connect TP to TP.
  - Function: Resist lateral flexion of the spine

Landmarks

Cervical Region

- **Anterior:**
  - C2: TP palpated at the angle of the mandible
  - C3: Hyoid bone
  - C4, C5: Thyroid cartilage
  - C6: First cricoid ring, carotid tubercle

- **Posterior:**
  - C2: First palpable midline spinous process (two fingerbreadths below the occiput)
  - C7: Vertebral prominens (largest cervical spinous process; nonbifid)
  - Articular pillars: Lateral off the spinous process bilaterally

Thoracic Region

- T3: Spine of the scapula
- T8: Inferior angle of the scapula
- T10: Umbilicus
- T12: Lowest rib

Lumbar Region

- L4: Iliac crests
- S2: PSIS

Back Musculature (Figure 4–158)

Extrinsic Back Muscles

- Superficial layer:
  - Trapezius
  - Latissimus dorsi
- Intermediate layer:
  - Serratus posterior superior and inferior

Intrinsic Back Muscles

- Superficial layer: Splenius capitis and cervicis
- Intermediate layer:
  - Erector spinae:
    - Iliocostalis: Lumborum, thoracis, cervicis
    - Longissimus: Thoracis, cervicis, capitis
    - Spinalis: Thoracis, cervicis, capitis
- Deep layer:
  - Transversospinalis muscles:
    - Semispinalis: Thoracis, cervicis, capitis
    - Multifidus
    - Rotators
  - Interspinalis, intertransversarii muscles

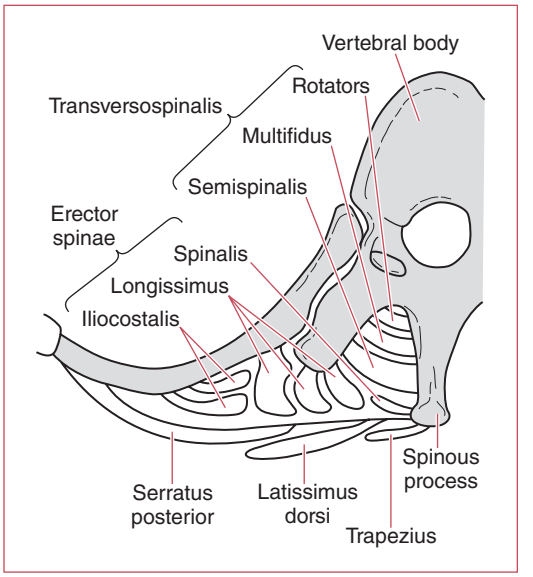

FIGURE 4–158 The back muscles: Transverse section, thoracic region.

Pertinent Spinal Biomechanics

POSITION	SPINE FLEXION
Erect posture	Mild activity in the erector spinae muscles
Initial flexion phase	Increased activity in the erector spinae muscles
Mid flexion phase	Increased activity in the gluteus maximus
Late flexion phase	Increased activity in the hamstrings
Terminal flexion phase	Electrical silence in erector spinae

JOINT	CERVICAL JOINT RANGE OF MOTION
AO joint	50% of flexion and extension of the entire cervical spine
AA joint	50% rotation of the entire cervical spine
C3–C7 joints	The remaining motion is distributed over the typical cervical segments

AA, atlanto-axial; AO, atlanto-occipital.

PATHOPHYSIOLOGY OF BACK PAIN

The Degenerative Cascade: Spondylosis (Figure 4–159)

General

- Kirkaldy-Willis and Burton (1992) presented a functional degenerative classification of the three-joint complex. It is initiated by a rotational strain or compressive force to the spine during lumbar flexion.
- This cascade consists of three phases: (a) dysfunction, (b) instability, and (c) stabilization; however, initial symptoms may present at any phase.
- Pathology of one component (disc or facet) influences deterioration of the other components (facet or disc) and adjacent vertebral level.
- This overall degeneration of the spine may be referred to as *spondylosis*.

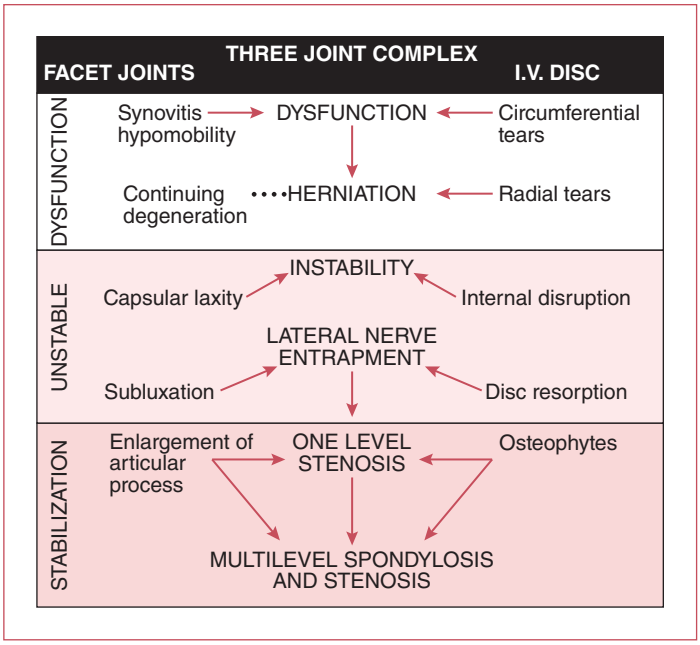

FIGURE 4–159 The degenerative cascade.

Phase

Dysfunction	This initial stage is typically a result of repetitive trauma. However, the pathologic changes that occur can be reversible. The Z-joints suffer minor capsular tears, cartilage degeneration, and synovitis, all of which lead to abnormal motion. The disc may have small annular tears and/or endplate separation. The segmental spinal muscles become hypertonic, splinting the spine and resulting in hypomobility.
Instability	Due to scar formation, each successive injury causes incomplete healing of the Z-joint capsules and annular fibers. With increased dysfunction, the joints have further degeneration of cartilage and increased capsular stretching/laxity. Annular disc tears progress with loss of nuclear substance. Overall, this results in hypermobility of the segments.
Stabilization	Progression leads to Z-joint articular cartilage destruction/hypertrophy, erosion, locking, and periarticular fibrosis. Discs have further loss of nuclear material, vertebral body approximation, endplate destruction, fibrosis, and osteophyte formation. Ankylosis can occur at the motion segment as well, entrapping spinal nerves. The patient may have an overall feeling of spinal stiffness.

DISC DISORDERS

DISC HERNIATION

General

- A herniated nucleus pulposus (HNP) is a disc injury in which the nuclear pulposus migrates through the annular fibers (annular tear), beyond the limits of the disc space margin. It may also initiate the release of enzyme phospholipase A2, which activates inflammatory mediators, such as leukotrienes, prostaglandins, platelet-activating factors, bradykinins, and cytokines.
- The disc material may include the nucleus, cartilage, bone, and anulus.

- The disc space margin is defined by the boundaries of the endplate and vertebrae.
- This most commonly happens at 30 to 40 years of age.
- A higher prevalence occurs for the lumbar region at the L4–L5 or L5–S1 discs followed by the C5–C6 disc.
- Fortunately, approximately three-fourths of these injuries will resolve with conservative care in 6 months to 1 year.

CLASSIFICATION (FIGURE 4–160)

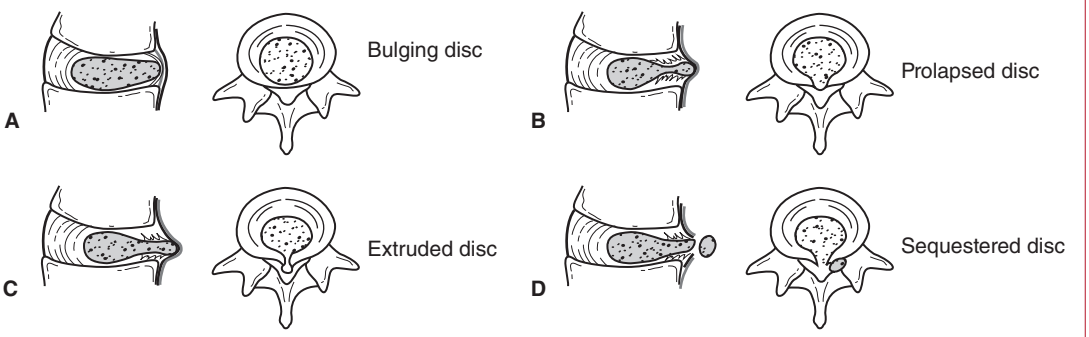

FIGURE 4–160 Disc classifications. (A) Bulging disc. (B) Prolapsed disc. (C) Extruded disc. (D) Sequestered disc.

TRADITIONAL DISC CLASSIFICATION	
A. Bulging disc	No annulus defect. Disc convexity is beyond vertebral margins (borders).
B. Prolapsed disc	Nuclear material protrudes into an annulus defect.
C. Extruded disc	Nuclear material extends to the posterior longitudinal ligament.
D. Sequestered disc	Nuclear fragment free in the canal.

Disc Herniation Location (Figure 4–161)

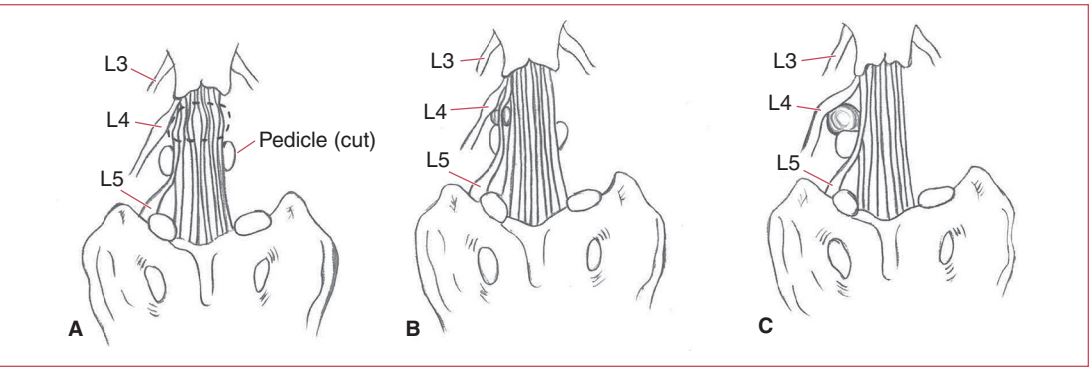

FIGURE 4–161 Disc herniation locations: (A) Central. (B) Posterolateral. (C) Far lateral.

Central	May present with axial spinal pain with or without radicular symptoms. Possible multi-root involvement if the cauda equina is affected or myelopathy if the spinal cord is involved.
Posterolateral	More common in the lumbar spine due to tapering presentation of the posterior longitudinal ligament; for example, a posterolateral L4–L5 herniation can impinge the L5 nerve root.
Lateral/ foraminal	May present with axial spinal pain with or without radicular symptoms. Affects the exiting root of that intervertebral level; for example, a lateral L4–L5 herniation can impinge the L4 nerve root.

Etiology

- Spontaneous
- Lifting activities
- Coughing/sneezing
- Bending/twisting activities

Clinical Features

- Symptoms depend on herniation location.
- Acute neck or back pain with or without radiating symptoms into the upper or lower limbs.
- Weakness, numbness, paresthesias, or pain secondary to chemical or mechanical stimuli to the disc, or nerve root irritation. A lateral lumbar list or shift may be noted.
- Exacerbation occurs with lumbar motion (forward flexion: central and posterior lateral HNP; extension: lateral HNP), sitting, sneezing, coughing, or Valsalva maneuvers, as well as neural tension tests.

Dermatomal and Peripheral Nerve Distribution (Figure 4–162)

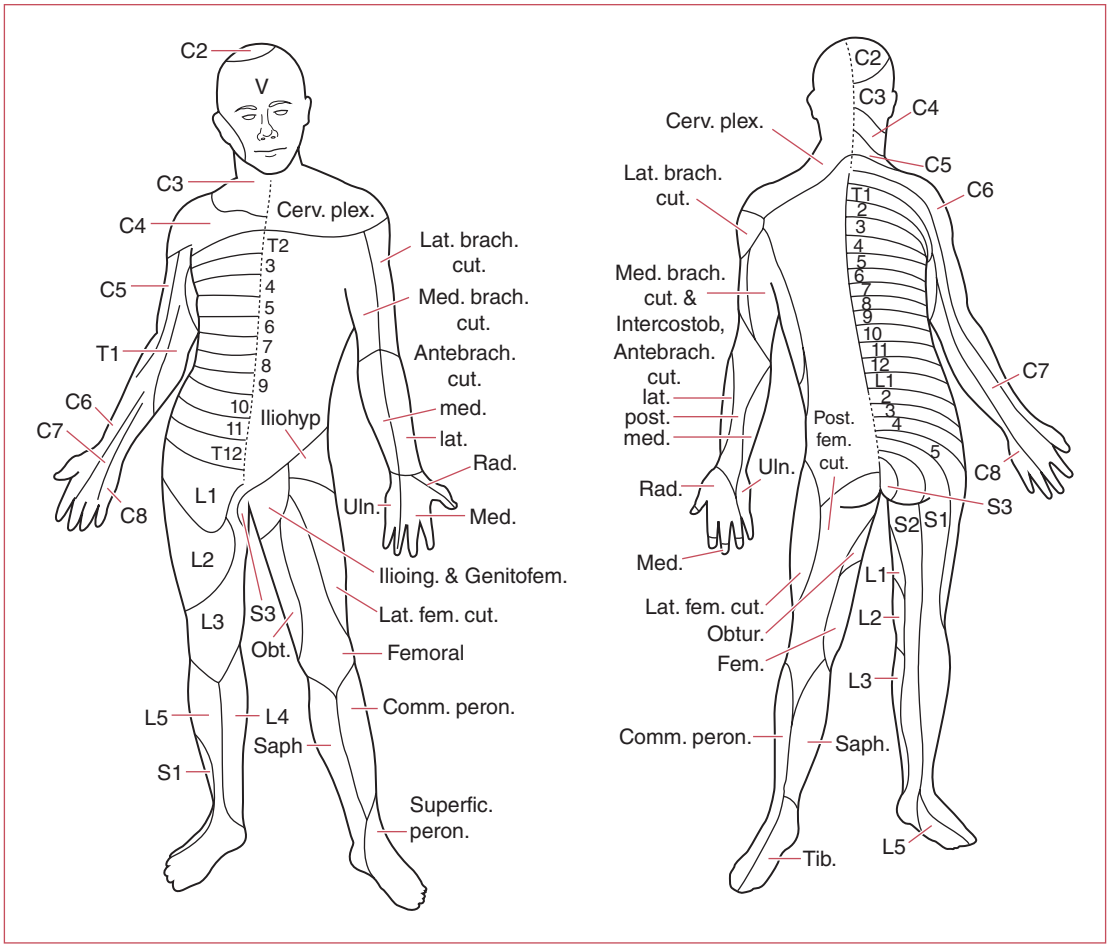

FIGURE 4–162 Dermatome and peripheral nerve distribution. Note: The common peroneal nerve (Comm. peron. in the figure) is also called the common fibular nerve. The superficial peroneal nerve (Superfic. peron. in the figure) is also called the superficial fibular nerve. The lateral femoral cutaneous nerve (Lat. fem. cut. in the figure) is also called the lateral cutaneous nerve of the thigh.

ROOT	MUSCLE WEAKNESS	REFLEX ABNORMALITIES	SENSORY DEFICITS
C5	Biceps brachii	Biceps brachii	Lateral arm
C6	Extensor carpi radialis	Brachioradialis	Lateral forearm
C7	Triceps brachii	Triceps brachii	Middle finger
C8	Flexor digitorum profundus	None	Medial forearm
T1	ADM/Interossei	None	Medial arm
T2, T4	None	None	• Band-like presentation based on segmental innervation • Lack of abdominal or chest pain T2—apex of axilla T4—nipple line T6—xiphoid process T10—umbilicus T12—inguinal ligament
T5–T10	Upper rectus abdominus		
T10–L1	Lower rectus abdominus		
L2	Iliopsoas	Cremasteric	Anterior thigh
L3	Quadriceps	Patellar	Anterior and lateral thigh
L4	Tibialis anterior	Patellar	Medial malleolus
L5	Extensor hallucis longus	Medial hamstring	Dorsum of the foot
S1	Gastrocnemius-soleus	Achilles	Lateral foot and little toe

ADM, abductor digiti minimi.

PROVOCATIVE TESTS FOR RADICULOPATHY

CERVICAL SPINE

Spurling's Test (Figure 4–163)

- Reproduction of radicular symptoms with cervical spine extension, rotation, and lateral flexion of the seated patient

FIGURE 4–163 Spurling's test.
Source: Courtesy of JFK Johnson Rehabilitation Institute. George Higgins, 2000.

Cervical Compression Test (Figure 4–164)

- Reproduction of radicular symptoms with a downward compression on top of the head

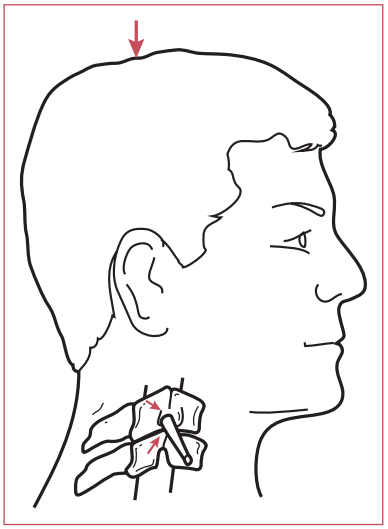

FIGURE 4–164 Cervical compression test.

(*continued*)

LUMBAR SPINE

Straight Leg Raise Test (Lasegue's Test; Figure 4–165)

- Reproduction of radicular symptoms with passive hip flexion of the extended leg while the patient is lying supine. This creates sciatic nerve tension at 30–60 degrees.
- Test sensitivity may be increased with dorsiflexion of the ankle (Lasegue's sign).
- A crossed straight leg raise test reproduces pain on the involved side with flexion of the opposite hip.

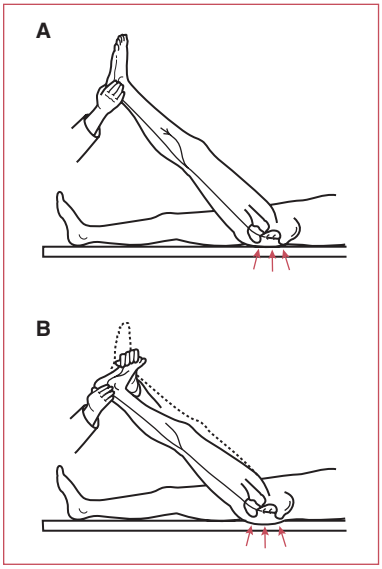

FIGURE 4–165 Straight leg raise test. (A) Sciatic nerve tension at 30 to 60 degrees of hip flexion. (B) Dorsiflexion of ankle may produce Lasegue's sign.

Bowstring Test (Figure 4–166)

- After a positive straight leg raise is elicited, decrease the angle of hip elevation to decrease the radicular pain. Then add pressure to the popliteal fossa over the nerve to reproduce symptoms.

FIGURE 4–166 Bowstring test.
Source: Courtesy of JFK Johnson Rehabilitation Institute, 2000.

Femoral Nerve Stretch Test (Reverse SLR or Ely's Test; Figure 4–167)

- Reproduction of anterior thigh pain in the prone patient with knee flexion and hip extension. This will stretch the femoral nerve and L2–L4 roots.

FIGURE 4–167 Femoral stretch test.
Source: Courtesy of JFK Johnson Rehabilitation Institute, 2000.

Sitting Root Test (Figure 4–168)

- Reproduction of radicular symptoms with a seated patient in a slumped posture, with cervical spine flexion and knee extension.

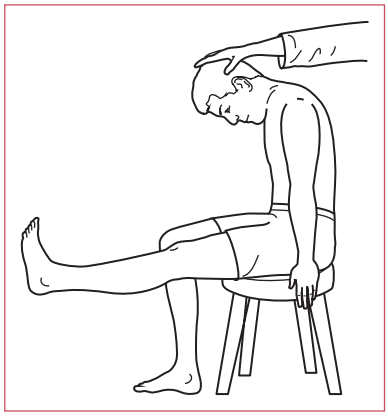

FIGURE 4–168 Slump test.

(*continued*)

UPPER MOTOR NEURON SIGNS

Plantar Responses (Babinski's Sign; Figure 4–169)

- Rub the sole of the foot from a lateral to medial direction up the arch (hindfoot to forefoot direction), and monitor for an upgoing great toe.

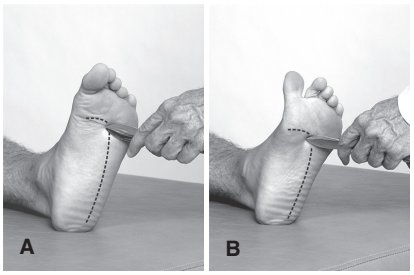

FIGURE 4–169 Babinski test—plantar responses. (A) Negative. (B) Positive. *Source:* Courtesy of JFK Johnson Rehabilitation Institute, 2000.

Hoffmann's Sign (Figure 4–170)

- Flick the patient's extended middle finger and monitor for twitching of the thumb and pointer finger.

FIGURE 4–170 Hoffmann's sign. *Source:* Courtesy of JFK Johnson Rehabilitation Institute, 2000.

SLR, straight leg raise.

Diagnostic Studies

- Imaging: X-rays, MRI, CT myelogram
- X-ray findings: Decreased disc height, vertebral osteophytosis and sclerosis, foraminal narrowing, facet arthrosis
- MRI: Demonstrates soft tissue pathology better than x-ray or CT, such as disc desiccation, annular tears, disc herniations, and nerve impingements
- MRI is the most common study to diagnose a herniated disc. Different structures demonstrate classic characteristics in appearance based on spin sequence. (Quick reminder—water [H_2O] is white on a T2 image. H_2O = T2.
- 🕮 Contrast-enhanced MRI: While the use of IV Gadolinium is not without potential complications, contrast-enhanced MRIs help differentiate postoperative scar tissue from recurrent disc herniation.

Treatment

- Disc herniations may regress on their own. Symptoms may resolve with or without changes in the mass effect of the herniation.
- *Conservative:*
  - Relative rest: Strict bedrest is NOT recommended.
  - Medications:
    - NSAIDs, analgesics, oral steroids, adjuvants (tricyclic antidepressants, selective serotonin reuptake inhibitors), muscle relaxants.
  - Rehabilitation program:
    - Patient education
    - Stretching program with a focus on hamstring flexibility
    - Strengthening program with a focus on core strengthening exercises
    - Spinal stabilization:
      - McKenzie program is a physical therapy program with a directional bias designed to centralize extremity pain.
      - Extension-biased programs may be used for posterior lateral HNP.
      - Neutral or flexion-biased program may be used for far-lateral HNP.
    - Modalities:
      - Thermal therapies (heat, cold), electric stimulation
      - Traction:

    - Vertebral distraction may relieve nerve compression:
        - 🕮 Cervical region: 20 to 30 degrees of flexion with 25 lb. of resistance. Less flexion is required for treatment of muscle spasm.
        - 🕮 Lumbar region: May require increased force or a split table to overcome friction
    - Indications:
        - Radicular pain (most widely accepted)
        - Paraspinal muscle spasm
    - Contraindications:
        - Ligamentous instability
        - Radiculopathy of uncertain etiology
        - Acute injury
        - RA
        - Vertebrobasilar arteriosclerotic disease
        - Spinal infections (Pott's disease)
  - Bracing: Lumbar corset for comfort. (Note: Abdominal/trunk musculature weakness may occur with prolonged bracing due to disuse atrophy.)
  - Home exercise program
  - Psychological interventions, muscle relaxation techniques, acupuncture
- Epidural steroid injections (ESIs):
  - Also see section Interventional Spinal Procedures in Chapter 11, "Pain Medicine," for a more detailed discussion.
  - Should be performed under fluoroscopic guidance with contrast-enhanced techniques.
  - Mechanism: Decrease inflammation causing nerve-root irritation through corticosteroid injection of the specific nerve root(s).
  - Complications/side effects:
    - Needle placement: Bleeding, infection, tissue damage, nerve injuries including spinal cord injury (Ziai et al., 2006)
    - Anesthetic: Confusion, anaphylaxis, convulsions, seizures, or death with intravascular injection
    - Corticosteroid: Immunosuppression, fluid and electrolyte imbalance, adrenal suppression, symptom flare. Exacerbation of underlying medical conditions: Diabetes, congestive heart failure, or hypertension.
    - Others, for example, stroke from embolus
- 🕮 Chymopapain injections:
  - Mechanism: Dissolve subligamentous herniations contained by the PLL.
  - Complication/side effects: Anaphylactic reaction, chronic pain, poor efficacy
- *Surgical intervention:*
  - May demonstrate quicker initial resolution of radicular pain but has not shown to have any greater statistical advantage over conservative measures with time
  - Surgery may improve leg pain. However, after 2 years, function and back pain are the same as with conservative care (Weinstein et al., 2006).
  - Considered for unremitting pain unresponsive to more conservative treatments, progressive weakness, cauda equina syndrome, or myelopathy

CAUDA EQUINA SYNDROME (FIGURE 4–171; ALSO SEE CHAPTER 7, SPINAL CORD INJURIES)

General

- Injury to the nerve roots forming the cauda equina
- Usually a result of a large central disc herniation
- Other causes include spinal stenosis, epidural tumors, hematomas, abscesses, and trauma.

Clinical Features

- Lumbar, buttock, perianal discomfort, and lower limb weakness
- Lower motor neurons signs including hyporeflexia

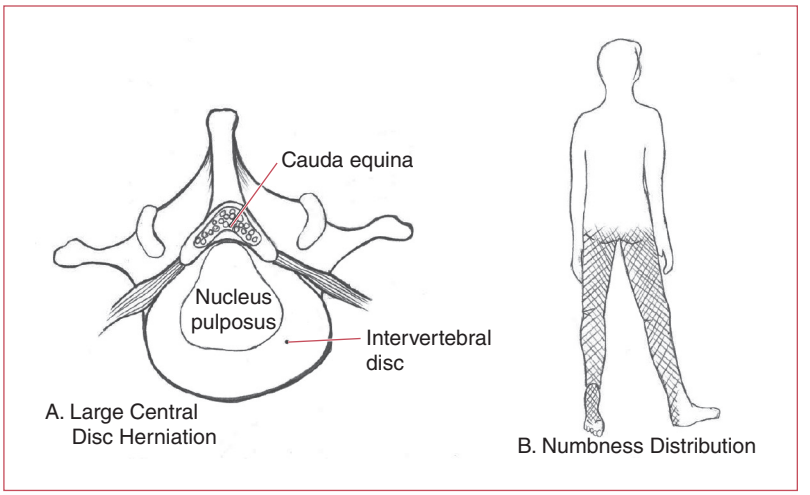

FIGURE 4–171 Cauda equina syndrome.

- Neurogenic bowel and bladder abnormalities (retention, frequency, incontinence)
- Sexual dysfunction
- Saddle anesthesia including the back of the legs, buttocks, and soles of the feet

MYELOPATHY

General

- 🕮 Injury to the spinal cord. Patients can have a history of radiculopathy, disc herniation, or stenosis. Tumors, arteriovenous (AV) malformations, multiple sclerosis, syphilis, syringomyelia, amyotrophic lateral sclerosis, spinal instability, or RA (C1–C2 subluxation) may also be considered.

Clinical Features

- Spastic or ataxic gait abnormalities, weakness, sensory changes, neurogenic bowel and/or bladder.
- Upper motor neuron signs include hyperreflexia, clonus, spasticity, Lhermitte's sign, positive Babinski's and Hoffmann's signs.

INTERNAL DISC DISRUPTION

General

- This is the degradation of the internal architecture of the disc without a gross herniation. It is associated with annular fissures and nuclear tissue disorganization. The degradation of nuclear material can lead to radial fissures and erosion of the annulus, causing chemical and mechanical stimulation of nociceptive fibers.

GRADING OF INTERNAL DISC DISRUPTION (FIGURE 4–172)	
0	No annular disruption
1	Inner one-third annular disruption
2	Inner two-thirds annular disruption
3	Outer one-third annular disruption ± circumferential spreading

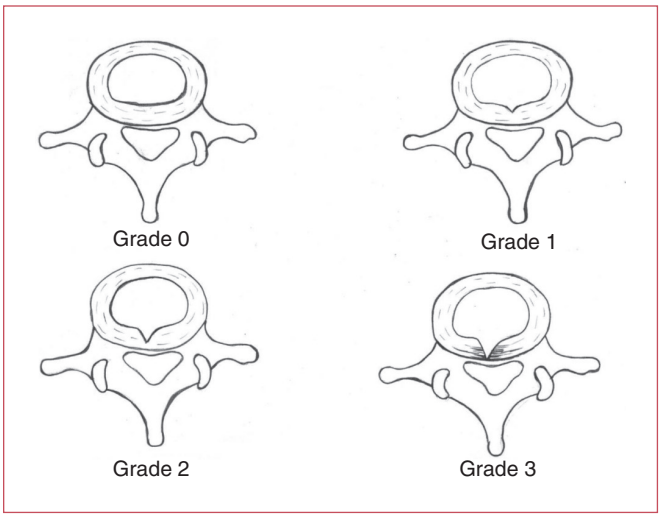

FIGURE 4–172 Internal disc disruption. Grades of radial fissures in internal disc disruption. (See text for description of grades.)

Etiology (Figure 4–173)

- Endplate fractures from excessive loads
- 🕮 **Risk Factors:** For intervertebral disc pathology range from environmental (behavioral, mechanical) to genetic influences.

Environmental Factors

- Weight: Excess body weight increases the chance for a herniated disc and poses a 12× increased chance to reherniate the same disc.
- Occupation: Physically demanding job activities. This includes heavy lifting, and repetitive pushing, pulling and twisting activities.
- Age: Increased typically between 35 and 50 years of age. This decreases after age 80.
- Sex: 2:1 male:female increased chance of a disc herniation.
- Smoking: Causes contraction of capillary vessels, with decreased nutritional diffusion. Nicotine inhibits cellular proliferation in the disc, accelerating disc degeneration and pathology.

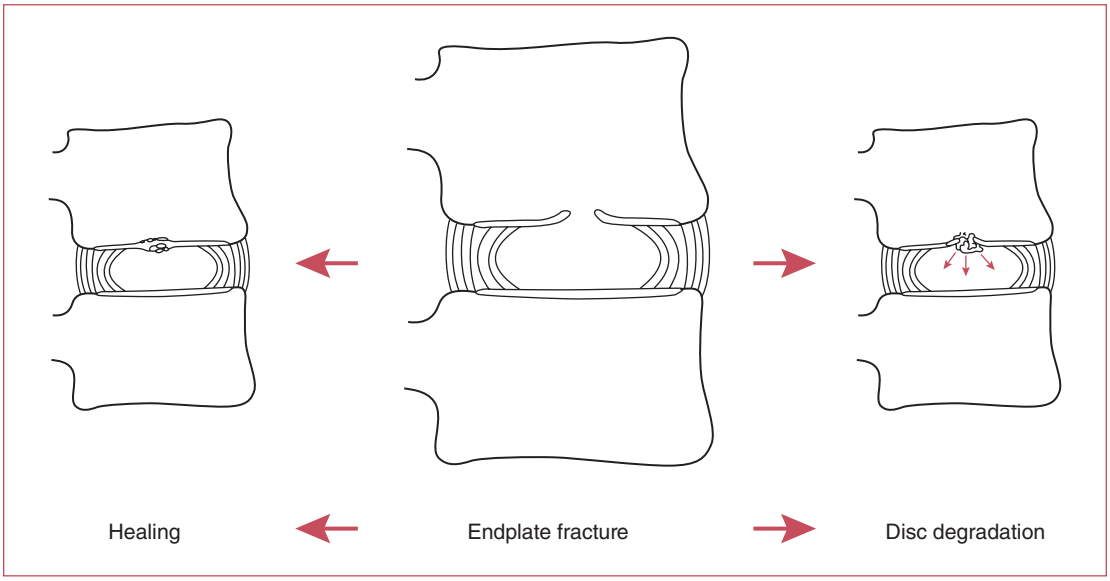

FIGURE 4–173 Possible outcome of endplate fractures: Compression of the intervertebral disc results in fracture of a vertebral endplate. The fracture may heal or trigger intervertebral disc degradation.

Genetics

- Hereditary predisposition to disc injury. Seen in twin study models regardless of environmental factors. This is influenced by vitamin D receptors, collagen synthesis mutations, and enzyme and cytokine production.

Clinical Features

- Constant, deep, aching axial discomfort, increased with mechanical stresses; that is, sitting, bending, twisting, lifting.
- May have absent neurologic involvement.

Imaging

- Imaging: MRI, CT, discogram
- Radial fissures are best demonstrated on post-discogram CT.
- A high-intensity zone in the annulus may be seen on T2-weighted MRI.

Treatment

- *Conservative:*
  - Relative rest, medications; rehabilitation program
  - ESIs may have potential benefit.
  - Intradiscal electrothermal therapy (IDET) annuloplasty has not been shown to be an effective treatment (Freeman, 2006).
- *Surgical:*
  - Spinal procedures including fusion stabilization may be considered for patients with unremitting pain if more conservative measures have failed.

BONE DISORDERS OF THE SPINE

SPINAL STENOSIS

General

- Degenerative changes occurring in the spine that result in disc space narrowing, vertebral body osteophytosis, facet joint arthropathy, and ligamentum flavum hypertrophy.
- These changes can cause stenosis of the central canal, lateral recess, or neuroforamina in the spine and can lead to nerve impingement.
- Neural compression or ischemia can cause associated limb pain syndromes that usually present at approximately 50 years of age.
- Involvement of the lumbar region is most common, affecting the L4–L5 levels.

Classification

- Central spinal stenosis (**Figure 4–174**):
  - Decreased size of the central spinal canal, typically secondary to spondylotic changes, including facet and ligamentum flavum hypertrophy. Can also occur due to other

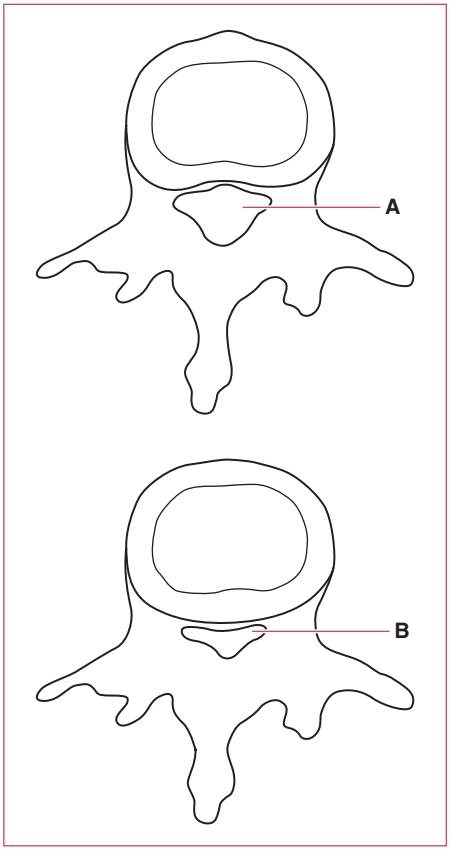

FIGURE 4–174 Central spinal stenosis. (A) Normal. (B) Central canal stenosis.

conditions including large disc herniation, epidural lipomatosis, epidural hematoma, or degenerative spondylolisthesis.
  - Cervical spine AP dimensions:
    - The normal spinal cord is approximately 10 mm in diameter; the spinal canal is 17 mm.
    - Neurologic sequelae may begin when the central canal is <12 mm (relative stenosis) to 10 mm (absolute stenosis).
- Lateral recess stenosis **(Figure 4–175)**:
  - Lumbar spine: Lateral stenosis has been further subdivided into three areas of entrapment, across the motion segment:
    - Lateral recess
    - Mid zone
    - Intervertebral foramen

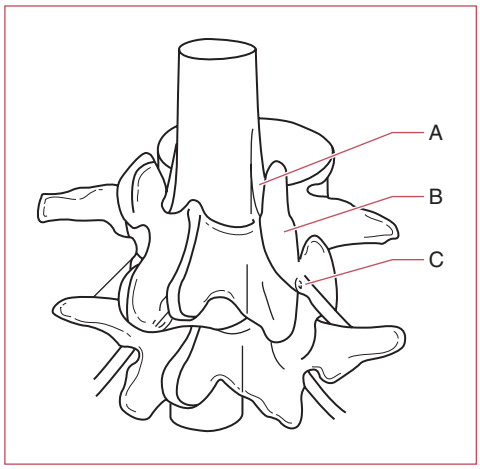

FIGURE 4–175 Lateral stenosis. (A) Lateral recess. (B) Mid zone. (C) Intervertebral foramen.

ZONES	ENTRANCE ZONE (LATERAL RECESS)	FORAMINAL STENOSIS: MID ZONE (PARS REGION)	FORAMINAL STENOSIS: EXIT ZONE (INTERVERTEBRAL FORAMEN)
Borders	Posterior: SAP Anterior: Posterior vertebral body and disc Medial and lateral walls: Open	Posterior: Pars interarticularis Anterior: Posterior vertebral body Medial wall: Open	Posterior: Z-joint (inferior level) Anterior: Posterior disc (inferior level)
Contents	Descending nerve root	Dorsal root ganglia Ventral motor root	Spinal nerve
Etiology	Hypertrophic facet joints	Osteophytes under the pars	Hypertrophic facet joints
Root Level	Nerve root exiting below (L3–L4 lateral recess involves L4 nerve root)	Same as the vertebrae (L3 pars involve L3 roots)	One level up from the vertebrae (L4 SAP or L3–L4 discs involve L3 roots)

SAP, superior articulate process.

Etiology

- Congenital:
  - Hereditary
  - Achondroplastic
- Acquired:
  - Degenerative (most common)
  - Spondylosis/spondylolisthesis
  - Iatrogenic (post-laminectomy/fusion)
  - Posttraumatic
  - Metabolic (Paget's disease)
  - Mass lesion (e.g., disc herniation, tumor, abscess)

Clinical Features of Central Spinal Stenosis

- Also known as neurogenic claudication (pseudoclaudication). Note that neurogenic claudication symptoms are position-dependent: Symptoms typically exacerbate with lumbar extension (standing/walking) and improved with lumbar flexion (sitting/bending forward). In contrast, vascular claudication symptoms change with the degree of exertion and not by position: Increased activity exacerbates symptoms (due to increased vascular demand from the leg muscles) when sitting or upright and improve with rest.

- Pain from neurogenic claudication may present with pain in the buttock, thigh, or leg with standing or walking, which is relieved with sitting or leaning forward ("shopping cart sign"). This may be due to neural compression due to structural compression or venous congestion.
- Gradual neck or back discomfort with upper or lower limb involvement
- Myelopathic changes may be noted with higher spinal cord level involvement if there is significant enough cord compression.

Imaging

- X-rays, MRI, CT, CT myelogram:
  - MRI is the ideal imaging study to identify central spinal stenosis. Findings typically include spondylotic changes, including vertebral body osteophytosis, ligamentum flavum hypertrophy, and facet joint arthropathy, which result in narrowing of the spinal canal.

Treatment

- *Conservative:*
  - Relative rest, medications
  - Rehabilitation program: Focus on a flexion- or neutral-biased spinal stabilization program, aquatic therapy.
  - ESIs
- *Surgical:*
  - Surgical procedures including decompression and/or stabilization

NEUROGENIC VERSUS VASCULAR CLAUDICATION

Symptoms	Neurogenic	Vascular
Discomfort	Numbness, aches, pain	Cramping, tightness
Location	Buttock, thigh, calf	Leg/calf
Exacerbation	Standing, walking, lying flat	Walking, cycling
Bicycle test	Painless	Painful
Downhill walking	Painful	Painless
Uphill walking	Painless	Painful
Remission	Flexed position, bending, sitting	Standing, resting, lying flat
Associated factors	Back pain, decreased spine motion, atrophy, weakness, normal pulses	Back pain rare, normal spine motion, rare atrophy or weakness, abnormal pulses, loss of hair, shiny skin

🕮 LUMBAR SPONDYLOSIS

General

- Spondylosis is the spinal degenerative process illustrated by radiographic changes involving the vertebral bodies, discs, and facet joints. Prevalence increases with age; however, it is not pathognomonic for increased pain syndromes. Related to the Degenerative Cascade.

Etiology

- Cumulative stressors
- Repetitive microtrauma

Clinical Features

- Asymptomatic
- Associated conditions may cause secondary conditions of instability, joint arthropathy, and spinal stenosis, contributing to neurogenic claudication and spinal nerve impingement, which may be apparent on electrodiagnostic studies in extreme progression.

Risk Factors

- Degenerative disc changes
- Abnormal biomechanics
- History of injury
- Facet abnormality
- Genetics

Diagnostic Studies

- Imaging: x-ray, MRI, and CT demonstrate associated spondylosis changes but are not necessarily associated with symptoms.
- Diagnostic injections can demonstrate supportive evidence of joint or nerve symptomatology regarding imaging findings.
- EMG testing can be considered to further evaluate radicular symptoms, if present.

Treatment

- Conservative
  - Care is primarily focused on the associated symptoms attributed to the pertinent clinical pathology.
  - Rehabilitation program
  - Interventional procedures
- Surgical
  - For secondary instability causing neurologic compromise

SPONDYLOLYSIS (FIGURES 4–176 AND 4–177)

General

- Vertebral defect most commonly seen in children and adolescents at the L5 vertebral level

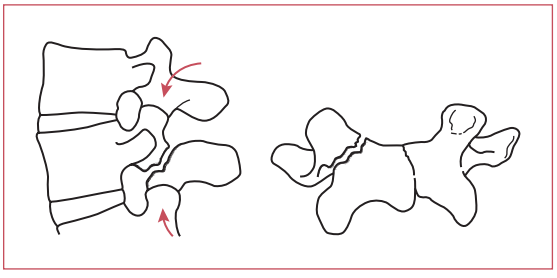

FIGURE 4–176 Lumbar spondylolysis.

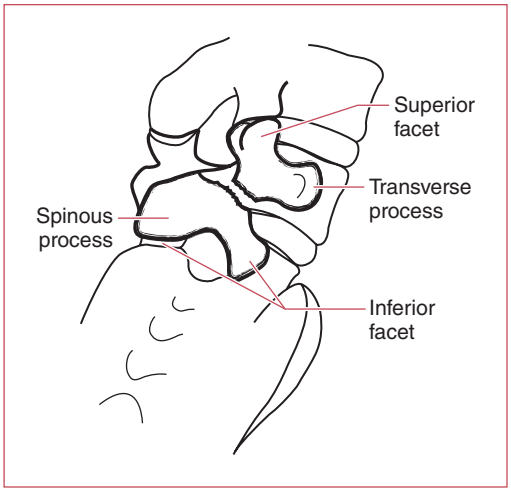

FIGURE 4–177 Spondylolysis (pars fracture of the neck of the "Scotty dog").

- It is a fracture of the pars interarticularis, which is located at the junction of the pedicle, lamina, and the SAP.
- This can lead to a spondylolisthesis (see the Spondylolisthesis section).

Etiology

- Most common forms occur as a result of repetitive hyperextension and rotational forces (e.g., sports such as gymnastics, football, wrestling) in young athletes.

Clinical Features

- Localized back pain exacerbated by motion (hyperextension), standing, lying prone, and relieved with flexion
- Neurologic exam should be normal.

Imaging

- X-rays: Oblique x-rays demonstrate a pars defect represented by a "broken neck" on the "Scotty dog" **(**Figure 4–176**)**.
- If x-rays are negative, bone scan can be considered:
  - Bone scans may be positive at 5 to 7 days and last up to 18 months.
  - Can help to differentiate between acute and chronic fractures
- If x-ray and bone scans are negative, and an acute pars fracture is still suspected, single-photon emission CT (SPECT) scan can be performed:
  - SPECT increases bone scanning sensitivity.
- High-resolution MRI may also be used.

Treatment

- Relative rest, avoidance of high-impact exercises/activities until pain-free, physical therapy, gradual transition back to play

SPONDYLOLISTHESIS (FIGURE 4–178)

General

- Slippage of a vertebral body with respect to the vertebral body below it. It may be an anterior translation (anterolisthesis) or posterior translation (retrolisthesis).
- It can be present at any vertebral level but is most commonly seen in the lumbar region.
- Occurrence is two to four times greater in males.
- There is no significant association between findings of spondylolisthesis and lower back pain (Kalichman et al., 2009, 2010).

Classification

- The grading of a spondylolisthesis is based on the percentage displacement with respect to the vertebral body or sacrum below **(**Figure 4–179**)**.

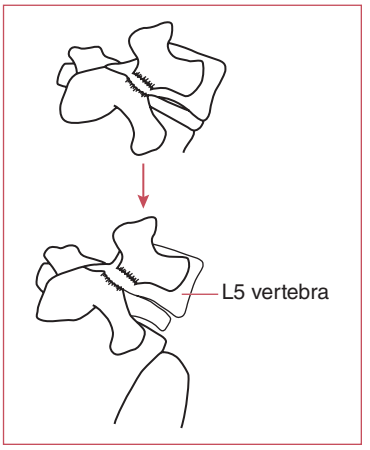

FIGURE 4–178 Spondylolysis with spondylolisthesis, oblique view. In this instance, the apparent collar (pars) on the Scotty dog widens because the vertebra has slipped.

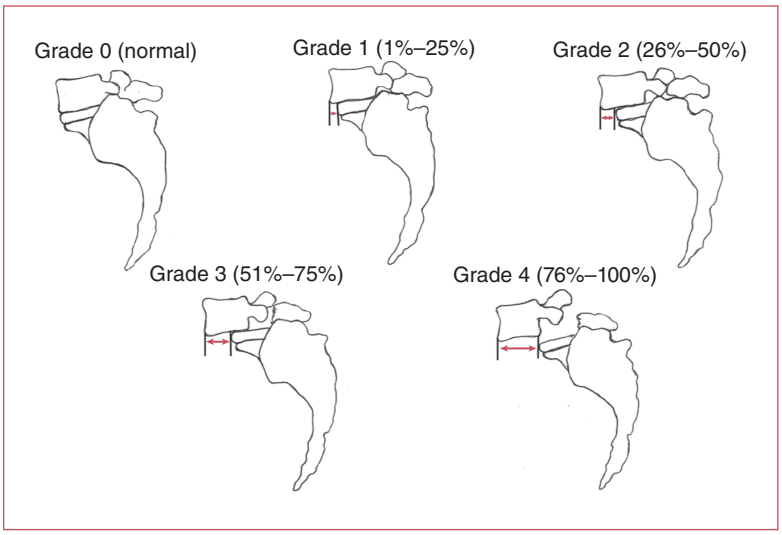

FIGURE 4–179 Meyerding grading of slippage (spondylolisthesis).

MEYERDING GRADING OF SLIPPAGE (SPONDYLOLISTHESIS)

Grade 0	0% slippage (normal)
Grade 1	1%–25% slippage
Grade 2	26%–50% slippage
Grade 3	51%–75% slippage
Grade 4	76%–100% slippage
Grade 5 Spondyloptosis	>100% slippage

Etiologies of Spondylolisthesis

CLASS	TYPE	AGE	CRITERIA
I	Dysplastic (congenital)	Child	Congenital abnormality of the lumbosacral Z-joint
II	Isthmic (most common type in adolescents and young adults)	5–50	Pars interarticularis fracture (subtype A), which is most common at L5–S1 or an elongation (subtype B)
III	Degenerative (most common type in adults)	Adult/ Elderly	Facet arthrosis causing subluxation; common location: L4–L5
IV	Traumatic	Young	Acute fracture in surrounding location other than the pars
V	Pathologic	Any	Generalized disease: Cancer, infection, metabolic disorder
VI	Postsurgical	Adult	Excessive resection of neural arches or facets causing an unstable structure

Clinical Features

- Low back pain exacerbated with motion, hamstring tightness, with palpable step-off noted at the slippage site
- Radicular symptoms may occur with marked slippage and result in central or foraminal stenosis.

Imaging

- X-rays, MRI, CT (Dreyfuss et al. 1995; Dreyer & Dreyfuss, 1996; Schwarzer et al., 1994):
  - Flexion and extension x-ray views may demonstrate signs of segmental instability.
- Radiographic instability:
  - Translation >3.5 mm (cervical) and >5 mm (thoracic or lumbar; **Figure 4–180**).
  - Rotational motion of two adjacent vertebrae >11 degrees in the C-spine and 15 degrees in the lumbar spine (**Figure 4–181**).

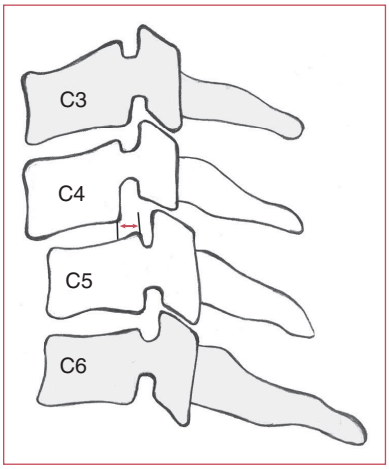

FIGURE 4–180 Increased cervical translation—sagittal plane. Cervical instability is translation >3.5 mm.

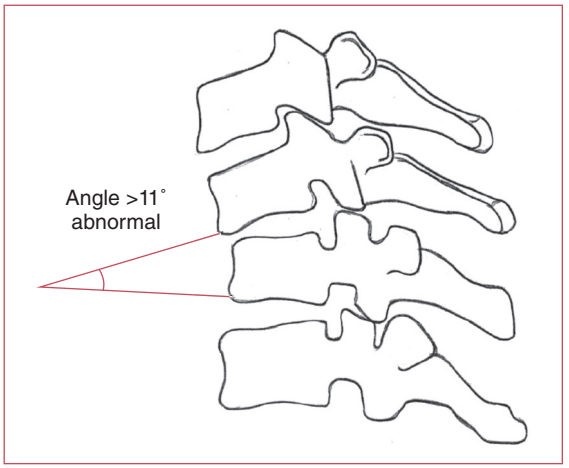

FIGURE 4–181 Increased cervical rotation—sagittal plane.

Treatment

- *Conservative:*
  - Grade 1, Grade 2, and asymptomatic Grade 3:
    - Relative rest, eliminate aggravating activities. Rehabilitation program: Focus on spinal stabilization exercises in a flexion-biased position and hamstring flexibility.
    - Asymptomatic Grade 1 spondylolisthesis may return to any activity but asymptomatic Grade 2 and 3 slips are restricted from contact sports. Progression of the spondylolisthesis is uncommon, and treatment will depend on risk factors and degree of angulation.
    - Thoracolumbosacral orthosis (TLSO) bracing is used if increased pain occurs despite decreased activity or an increase in slippage is suspected.
- *Surgical:*
  - Indicated in symptomatic Grade 3, Grades 4 and 5, unstable spondylolistheses, and refractory cases.
  - Surgery typically involves bilateral posterolateral fusion with or without decompression.

SCOLIOSIS (ALSO SEE CHAPTER 10, PEDIATRIC REHABILITATION)

General

- A general spinal deformity characterized by lateral curvature(s) and vertebral rotation.
- It may be associated with a fixed structural curve or reducible functional curve.
- Correlation with discomfort is unclear, but low back pain is usually the initial symptom. It is related to curve severity and usually begins at the convexity.

Etiology

	STRUCTURAL SCOLIOSIS	
Idiopathic	Most common	
	Subtypes	
	Infantile	Birth to 3 years: Associated with congenital defects
	Juvenile	4–10 years; high risk of curve progression
	Adolescent	Most common; 10 years to maturity; high risk of progression
Congenital	May be due to an early embryologic developmental defect	
	Subtypes	
	Open	Caused by myelomeningocele
	Closed	May be associated with neurologic deficits Associated with a wedged vertebra, hemivertebra, congenital bar, or block vertebrae
Neuromuscular	Certain neuromuscular disorders may have a rapid curve progression with associated pulmonary and spinal cord complications.	

Clinical Presentation

PATTERNS	CHARACTERISTICS
Right thoracic curve	Most common; the apex can typically be seen at T7 or T8
Double major curve (S-shaped curve)	Right thoracic with a left lumbar curve; little cosmetic deformity
Lumbar curve	Left lumbar curves greater than right lumbar curves
Thoracolumbar curve	Less cosmetic deformity than thoracic curve, may have rib and flank distortion
Left thoracic curve	Rare; may be associated with spinal cord abnormalities

Imaging

- Scoliosis survey x-rays help establish diagnosis and prognosis:
  - The need for follow-up scoliosis survey films will depend on skeletal maturity, patient age, and degree of curvature.
  - Younger patients with rapidly progressing curves warrant more frequent x-ray follow-up.
- Rotation (Figure 4–182):
  - Pedicle portion estimates the amount of vertebral rotation on the PA view.
  - Grading: 0 (no rotation) to 4 (complete pedicle rotation out of view).
- Curve: Cobb angle (Figure 4–183):
  - An angle formed by the perpendicular lines drawn from the endplates of the most tilted proximal and distal vertebrae to measure the scoliotic curve

Treatment

TREATMENT	DEGREE OF ANGULATION (COBB ANGLE)
Observation	<20 degrees
Bracing	20–40 degrees
Surgery	>40 degrees (>35 degrees for neuromuscular diseases)

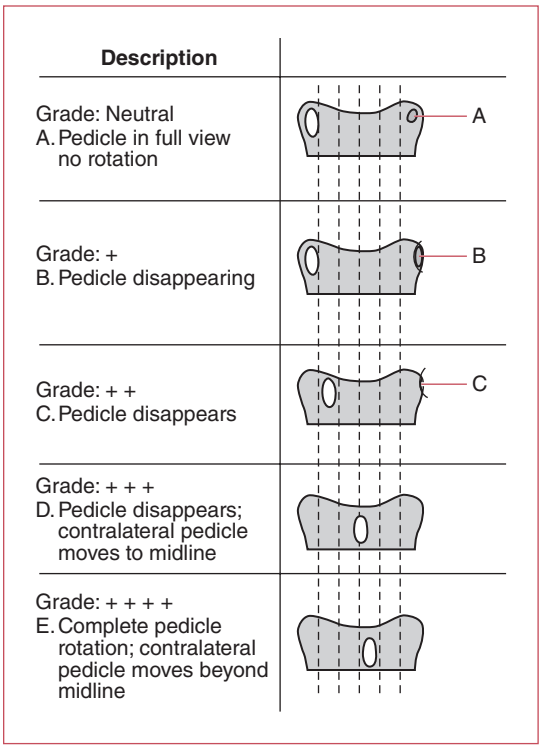

FIGURE 4–182 Measurement of vertebral rotation using pedicle method. Vertebral body is divided into six segments and grades from 0 to 4+ are assigned, depending on location of the pedicle within the segments.

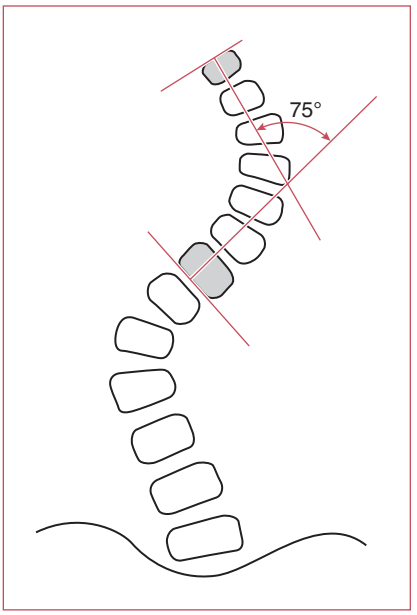

FIGURE 4–183 Cobb angle.

- *Conservative:*
  - Rehabilitation program
  - Bracing:
    - Prevents worsening of the curvature but does not correct scoliosis.
    - Worn 23 hours a day until spinal growth is completed.
    - Weaning off can begin when radiographs display signs of maturity and curves are stable.
    - Patients should be evaluated at 2- to 3-year intervals for life after the brace is discontinued.
  - *Types of bracing:*

Milwaukee brace	High thoracic curves (apex at T8)
Low-profile TLSO	Lower thoracic, thoracolumbar, and lumbar curves (apex below T8)

TLSO, thoracolumbosacral orthosis.

- *Surgical:*
  - Spinal surgical procedures are indicated for scoliosis with:
    - Relentless progression.
    - >40 degree curvature in the skeletally immature, >50 degrees in the skeletally mature, and >35 degrees in neuromuscular diseases, or progressive loss of pulmonary function.

SCHEUERMANN'S DISEASE (JUVENILE KYPHOSIS)

General

- 🕮 An adolescent disorder of the vertebral endplates and apophysis resulting in an increased thoracic kyphosis
- The kyphosis usually involves three sequential vertebrae and is generally >45 degrees.

Etiology

- Failure of endochondral ossification causing the following:
  - Intervertebral disc herniation.
  - Anterior wedging of the vertebral bodies in three or more adjacent levels.
  - Fixed thoracolumbar kyphosis.
- Recent literature also notes an autosomal dominant inheritance pattern (Bezalel et al., 2014).

Clinical Features

- More common in adolescent males.
- Can present with a progressive, nonpainful thoracic kyphosis.
- The thoracic kyphosis remains fixed and does not correct with hyperextension.
- Back pain may occur in young athletes due to localized stress injury to the vertebral growth plates.

Imaging

- Imaging: x-rays, CT, MRI
  - Anterior vertebral body wedging ≥5 degrees at three or more adjacent levels, irregular endplate, Schmorl's nodes with increased kyphosis angulation spanning three levels or more typically seen (Sardar et al., 2019).
  - Schmorl's nodes is a herniation of disc material through the vertebral endplate into the spongiosa of the vertebral body, and vertebral wedging of approximately 5 degrees (Figure 4–184)

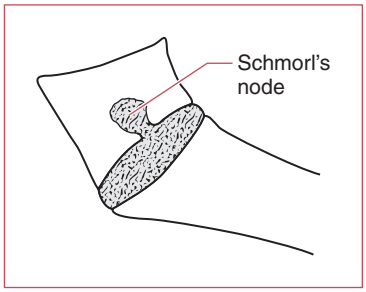

FIGURE 4–184 Schmorl's node.

Treatment

- *Conservative:*
  - Rehabilitation program: Focus on thoracic extension and abdominal strengthening exercises
  - Bracing may be used for kyphosis ≤74 degrees for a length of time dependent on skeletal maturity.
- *Surgical:*
  - Correction may be indicated if kyphosis is >75 degrees, or >60 degrees but with pain not responsive to other nonsurgical treatments (Hogue, 2022).

VERTEBRAL BODY COMPRESSION FRACTURE

General

- Typically associated with osteoporosis, these fractures are most commonly seen at the thoracolumbar junction. This is due to the transition between the fixed rigid thoracic and the highly mobile lumbar vertebra (see also Osteoporosis section in Chapter 12, Associated Topics in Physical Medicine and Rehabilitation).
- Denis (1983) described a three-column model for evaluating thoracolumbar fractures and determining their stability (Figure 4–185). (Also read section on Cancer Rehabilitation in Chapter 9, Pulmonary, Cardiac, and Cancer Rehabilitation.)

COLUMN	COMPONENTS	STABILITY
Anterior	• Anterior longitudinal ligament • Anterior two-thirds of the vertebral body and annulus fibrosis	Stable
Middle	• Posterior longitudinal ligament • Posterior one-third of the vertebral body and annulus fibrosis	Unstable
Posterior	• Ligamentum flavum, supraspinous and infraspinous ligament • Posterior elements: Pedicles, facets, spinous process	Stable

GENANT GRADING SYSTEM FOR VERTEBRAL BODY DEFORMITY	
Grade 0	Normal
Grade 1	Mild: 20%–25% height decrease
Grade 2	Moderate: 25%–40% height decrease
Grade 3	Severe: >40% height decrease

Genant HK, Wu CY, van Kuijk C, Nevitt MC. Vertebral fracture assessment using a semiquantitative technique. *J Bone Miner Res*. 1993;8:1137–1148. doi:10.1002/jbmr.5650080915

Etiology and Risk Factors

- Trauma
- Osteoporosis/osteopenia
- Osteomalacia
- Medication-related (chronic corticosteroids, chemotherapy, radiation, proton pump inhibitors)
- Neoplasm—most commonly from bony metastasis (also see section on Cancer Rehabilitation in Chapter 9, Pulmonary, Cardiac, and Cancer Rehabilitation)
- Infection
- Hyperthyroidism
- Smoking, alcohol abuse
- Female

Clinical Features

- Sudden onset of constant thoracolumbar pain. Can be related to a fall or can be atraumatic.
- Exacerbated with Valsalva maneuvers, turning in bed, coughing, flexion, or incidental trauma such as stepping off a curb.

Imaging

- Imaging: x-rays, bone scan, SPECT, CT, MRI
- Anterior vertebral body wedging typically seen on imaging studies (**Figure 4–186**). Bone scan with SPECT may have increased sensitivity.

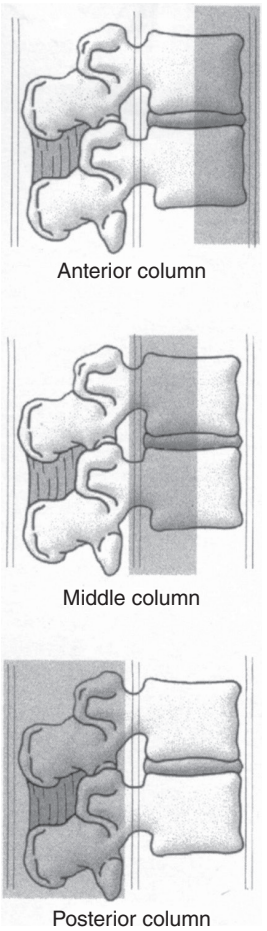

FIGURE 4–185 The three-column model of spinal stability. *Source:* From Nesathurai S, ed. *The Rehabilitation of People With Spinal Cord Injury: A House Officer's Guide*. Arbuckle Academic Publishers; 1999, with permission.

Treatment

- *Conservative:*
  - Indicated for fractures causing <25% decrease of vertebral height. Short-term bedrest followed by activity restriction.
  - Medications for pain control.
  - Rehabilitation program: Focus on hyperextension exercises.
  - Bone density evaluation if fracture is suspected to be due to bone loss.
  - Oncology evaluation if the fracture is suspected to be pathologic.
  - Bracing:

Elastic binder	• Acts as a reminder to limit motion • Increases intra-abdominal pressure
Custom-molded TLSO/Jewett brace	• Greater immobilization • Places patient in slight hyperflexion • Jewett should be used with caution in the osteoporotic patient

TLSO, thoracolumbosacral orthosis.

- *Interventional:*
  - Vertebral augmentation (vertebroplasty or kyphoplasty) is an option for cancer-related compression fractures when the symptoms are disabling with marked reduction in quality of life (Chandra et al., 2013).
  - Percutaneous vertebral augmentation has been increasingly utilized for osteoporotic fractures but is not considered a first-line treatment option. The long-term efficacy, cost-effectiveness, and safety of vertebroplasty and kyphoplasty remain unclear with the current literature. More recent, randomized controlled studies comparing vertebroplasty with sham showed no significant difference in outcomes for patients with osteoporotic vertebral body fractures. Additional studies are warranted to clarify its utility in treating osteoporotic fractures (Buchbinder et al., 2009; Kallmes et al., 2009; Wardlaw et al., 2009). It can be an option in refractory cases.

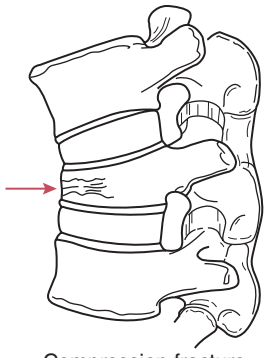

FIGURE 4–186 Thoracic compression fracture.

- *Surgical:*
  - Spinal procedures are indicated for fracture, causing >50% decrease of vertebral height, instability, and late kyphotic deformity leading to neurologic compromise.

VERTEBRAL BODY BURST FRACTURES

- Compression fractures of the vertebral body involving the anterior and middle columns of the spine from a significant trauma, typically from a fall from a height.
- Most commonly seen in the thoracolumbar region.
- Treatment is based on if it is stable or unstable:
  - Stable:
    - Neurologically intact. Posterior column remains intact.
    - <50% collapse of anterior vertebral body height.
  - Unstable:
    - Neurologic deficits present
    - >50% loss of anterior vertebral body height
    - Central canal compromise >30%
    - Posterior element injury.
- Radiographic findings:
  - X-ray, CT scan, MRI
- Treatment:
  - Stable: Typically treated nonoperatively with bracing for 4 to 6 months. Follow-up radiographs performed standing to evaluate for kyphosis.
  - Unstable: Surgical decompression and fusion to treat or avoid neurologic compromise.

JOINT DISORDERS OF THE SPINE

FACET SYNDROME

General

- A facet joint is a true synovial joint, containing a capsule, meniscus, and a synovial membrane.
- These joints also sustain progressively increasing compressive loads down the spine, reaching approximately 12% to 25% in the lumbar region.
- As the disc degenerates and decreases in height, greater loads are imparted on the joints and influence the degenerative cascade.

Etiology

- Somatic dysfunction/facilitated segment
- Positional overload
- Capsular tears/injury
- Meniscoid/synovial impingement
- Spondylosis pain patterns (posterior view).

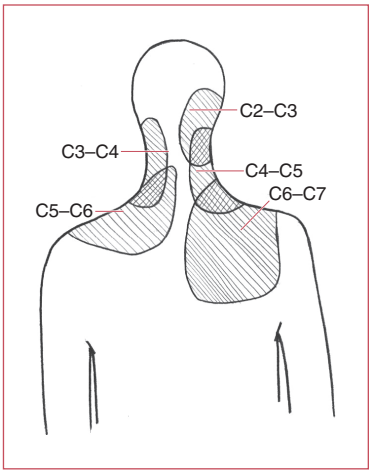

FIGURE 4–187 Cervical Z-joint referral pain patterns (posterior view).

Clinical Features

- Axial pain pattern with a radicular pain pattern presentation (**Figure 4–187**).
- Neck or back pain exacerbated with rotation and extension. No neurologic abnormalities.

Imaging

- Imaging: x-ray, MRI, CT:
  - No imaging study is specific for facet-mediated pain.
  - Degenerative changes may be noted but are not diagnostic.
  - MRI may show hypertrophy of the capsule and facets.
  - The gold standard in diagnosing facet pain is the use of double diagnostic medial branch blocks (Dreyfuss et al., 1995; Dreyer & Dreyfuss, 1996; Schwarzer et al., 1994).

Treatment

- *Conservative:*
  - Relative rest. Medications for pain control.
  - Rehabilitation program: Focus on spine stabilization in flexion-biased or neutral postures, proper body mechanics.
- *Interventional:*
  - Facet joint injections or dorsal rami medial branch radiofrequency ablation (RFA) only if diagnostic blocks are positive.
  - Surgical:
    - Endoscopic medial branch rhizotomy only if diagnostic medial branch block is effective.

SACROILIAC JOINT DYSFUNCTION

General

- The SI joint complex encompasses the SI joint capsule, the muscle and ligaments overlying the joint, and the lateral branch nerves innervating the joint anteriorly and posteriorly.
- 🕮 The SI joint is an ear-shaped articulation between the sacrum and the ilium that has a synovial joint anteriorly and syndesmosis posteriorly.
- It is innervated by the L4–L5 ventral rami, the superior gluteal nerve, and the dorsal rami of L5–S2 (Forst et al., 2006).
- There is a lack of a gold standard to diagnose SI joint pain.

Etiology

- Hyper/hypomobile joint patterns
- Repetitive overloads
- Trauma
- Capsular tears/injury

PROVOCATIVE SI JOINT TESTS

Patrick's Test (FABER; Figure 4–188)

- Pain reproduction with Flexion, ABduction, External Rotation of the hip joint, and Extension of the leg (downward force by the examiner). Ipsilateral pain occurs in a degenerative hip; contralateral pain occurs in the dysfunctional SI joint.

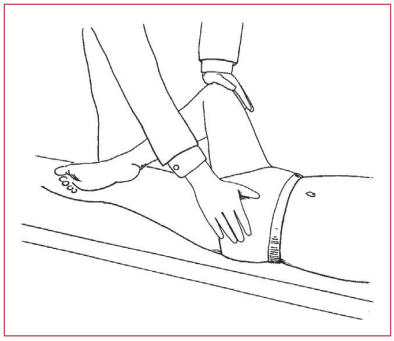

FIGURE 4–188 Patrick's (FABER) test.

Gaenslen's Test (Figure 4–189)

- SI joint pain is reproduced with extension of the involved leg off the table by the examiner while the contralateral hip is held in flexion.

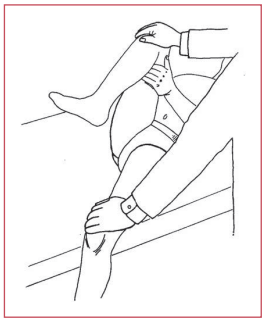

FIGURE 4–189 Gaenslen's test.

Iliac Compression Test (Figure 4–190)

SI joint pain with downward force placed on the iliac crest with the patient in a decubitus position

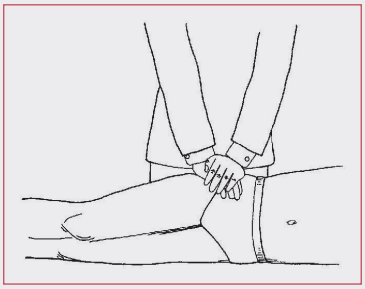

FIGURE 4–190 Iliac compression test.

SI, sacroiliac.

Clinical Features

- Acute or gradual back, buttock, leg, or groin pain with tenderness over the joint
- Abnormal SI joint motion patterns; increased discomfort with positional changes
- Discomfort within associated muscles, which may include the quadratus lumborum, erector spinae, and piriformis muscles
- No neurologic abnormalities are present.
- There is not a gold standard reference for clinical SI joint maneuvers. Clinical exam has significant false positive rates and variable sensitivity rates.

Imaging/Diagnostics

- X-ray, bone scan, CT, MRI
- These studies can be considered to rule out alternative pathologies in resistant cases. With the exception of sacroiliitis, imaging is unreliable in diagnosing SI joint dysfunction.
- Diagnostic SI joint blocks under fluoroscopic guidance have higher diagnostic value to diagnose SI joint pain.
- Serology workup can be indicated for underlying arthropathies.

NONPROVOCATIVE SI JOINT TESTS

Yeoman's Test (Figure 4–191)
SI joint pain with hip extension and ilium rotation

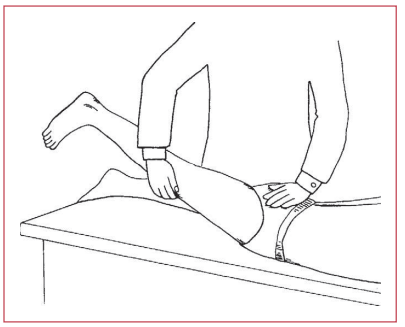

FIGURE 4–191 Yeoman's test.

Gillet's Test (Figure 4–192)

- Monitor PSIS motion when the patient raises the leg to 90 degrees. The PSIS on raised leg should rotate down. Restriction of this motion is considered abnormal.

FIGURE 4–192 Gillet's test.

Seated Flexion Test (Figure 4–193)

- Monitor the PSIS of the seated patient as they bend forward. Asymmetric cephalad motion of the PSIS indicates an SI dysfunction. Use the standing flexion test to distinguish the side of the dysfunction.

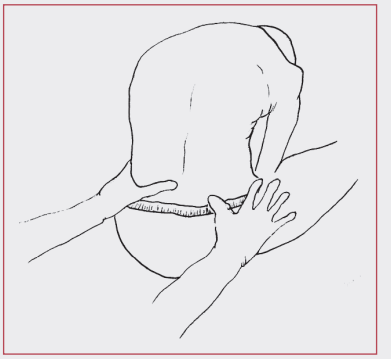

FIGURE 4–193 Seated flexion test.

PSIS, posterior superior iliac spine; SI, sacroiliac.

Treatment

- *Conservative*: Relative rest, medications. Rehabilitation program: Manual medicine, SI joint belt.
- *Interventional*: SI joint injections, RFA
- *Surgical*: Open or minimally invasive SI joint fusion, endoscopic lateral branch rhizotomy

SOFT TISSUE DISORDERS OF THE SPINE

🕮 SPRAIN/STRAIN

General

- This may be an overutilized term pertaining to muscular or ligamentous disruption due to overload injuries.

Etiology

- Overuse syndromes
- Excessive eccentric contraction
- Acceleration–deceleration injuries
- Acute trauma

Clinical Features

- Muscle aches with associated spasm and guarding in the region of injury.
- Delayed onset muscle soreness can occur within 24 to 48 hours typically after an eccentric overload injury.
- Facilitated segmental or somatic dysfunction may be more commonly involved than actual tissue disruption.
- Normal neurologic exam.

Imaging

- Decreased lordotic curvature may be seen on lateral x-rays, commonly due to muscle spasm.
- Imaging studies do not specifically diagnose a strain.

Treatment

- *Conservative*
  - Acute Phase (onset–1 month)
    - Pain control
    - Relative rest (prolonged bedrest not recommended), with return to pain-free activity when able.
    - Rehab program. ROM.
  - Subacute Phase (1–3 months)
    - Address proper body mechanics as activity level increases.
    - Core strengthening, ROM (hip flex/ext), and spine stability.
  - Chronic (>3 months)
    - Multidisciplinary approach: Therapies to improve strength and endurance; education to decrease future injuries; stress reduction; cognitive and behavioral therapy.
    - Evaluate for depression due to loss of physical abilities.

MYOFASCIAL PAIN SYNDROME (SEE ALSO "TRIGGER POINTS" SECTION)

General

- Denotes a regional pain disorder characterized by hypersensitive areas of taut muscle bands called myofascial trigger points
  - A trigger point is distinguished from a tender point by a circumscribed area of tenderness with a palpable, tense band of muscle fibers that causes concordant pain in a referred pain pattern with an associated local twitch response upon palpation (**Figure 4–194**).
- It may also cause decreased ROM and weakness due to the pain.

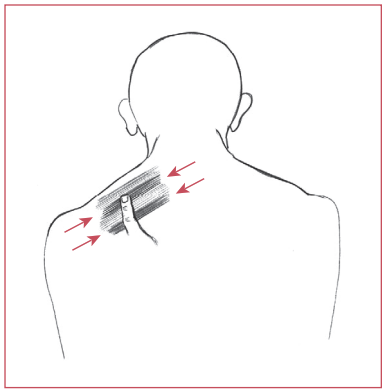

FIGURE 4–194 Myofascial trigger point: Pulling the taut band under the fingertip at the trigger point (dark stippled area) produces a "local twitch response" with shortening of the band of muscle.

Etiology

- Postural mechanics
- Overuse injuries
- Deconditioning
- Trauma
- Stress

Clinical Features

- Muscle tenderness, spasm, trigger points, decreased ROM.
- Nonmuscular symptoms including paresthesias, poor sleep patterns, and fatigue.
- Normal neurologic exam.

Imaging

- None available. Consider further workup if other potential pathologies are suspected.

Treatment

- *Conservative:*
  - Correct underlying causes. Analgesic medications PRN for discomfort.
  - Rehabilitation program focused on flexibility, strengthening, spine stabilization, and aerobic exercises.
  - Spray and stretch or trigger point injections may be beneficial.
  - Psychological counseling

FIBROMYALGIA

- See Chapter 3, Rheumatology and Chapter 11, Pain Medicine.

INFECTIONS OF THE SPINE

🕮 VERTEBRAL BODY OSTEOMYELITIS AND DISCITIS

General

- 🕮 An embolic infection of the vertebral body metaphysis causing ischemia, infarct, and bony destruction with disc involvement. Risk factors include advanced age, diabetes, immunodeficiency, penetrating trauma, dental infections, genitourinary (GU) procedures, and invasive spinal procedures. It is most commonly seen in the lumbar spine but increases in the cervical region with intravenous (IV) drug abuse and in the thoracolumbar junction with TB.

Etiology

- 🕮 *Staphylococcus aureus* (most common).
- *Pseudomonas* (IV drug abuse).
- 🕮 *Mycobacterium tuberculi* (Pott's disease) (**Figure 4–195**).

Clinical Features

- Fever and back pain.
- Potential spinal deformity if there is collapse of vertebral body(ies).
- Neurologic involvement including radicular pain, myelopathy, or paralysis can occur due to direct dural invasion with compression from an epidural abscess.
- Most commonly involves thoracic > lumbar spine.

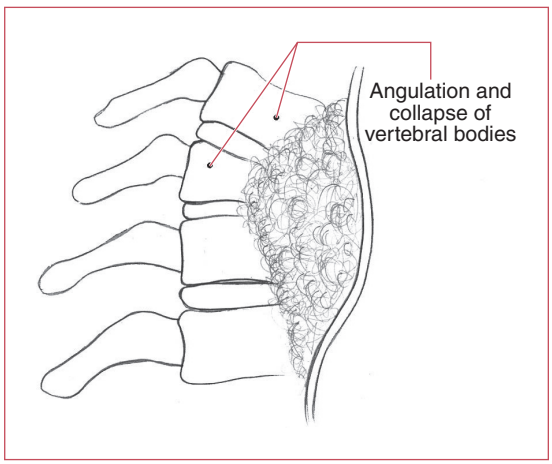

FIGURE 4–195 Pott's disease: Spinal tuberculosis.

Diagnostic Studies

- X-rays: By 2 weeks, radiographs can demonstrate evidence of endplate destruction.
- MRI is most sensitive and specific:
  - Findings include endplate erosion, discitis, osteomyelitis, and abscess.
  - T2: Hyperintense signal in vertebral body/disc.
  - T1: Hypointense signal in corresponding regions.
- Bone scan and SPECT
- CT shows hypodensity with trabecular, cortical, and endplate destruction.
- Lab work:
  - Leukocytosis, elevated erythrocyte sedimentation rate (ESR), and C-reactive protein (CRP).
  - Positive Gram stain and cultures.
- Positive bone biopsy.

Treatment

- *Conservative:*
  - IV and oral antibiotics:
    - *S. aureus*: Penicillin, first- or second-generation cephalosporins
    - *Pseudomonas*: Extended-spectrum penicillins
    - TB: 12 months mycobacterial agents (rifampin, isoniazid [INH], ethambutol, pyrazinamide)
  - Spinal immobilization with casting or bracing. Early ambulation.
- *Surgical:*
  - Spinal procedure including decompression and spine stabilization

ORGANIC NONSPINAL SOURCES OF BACK PAIN

General

- Factors causing spinal pain can be associated with other medical conditions. These disorders must be considered with any pain presentation as they can be the primary dysfunction, though the predominating symptom appears spinal.

Etiology

Visceral disorders	GU (prostatitis, renolithiasis, UTI), GYN (endometriosis, PID, ectopic pregnancy), GI (pancreatitis, cholecystitis, PUD)
Psychological disorders	Depression, anxiety, hysteria, somatization disorders
Neoplastic disorders	Primary tumors, metastatic tumors, multiple myeloma, lymphoma, leukemia, retroperitoneal tumor
Vascular disorders	Aortic aneurysm (back pain associated with pulsatile abdominal mass)
Rheumatologic disorders	Seronegative spondyloarthropathies (i.e., ankylosing spondylitis, psoriatic spondylitis, Reiter's syndrome, inflammatory bowel disease) and Paget's disease
Hematologic disorders	Sickle cell anemia, thalassemia

GI, gastrointestinal; GU, genitourinary; GYN, gynecologic; PID, pelvic inflammatory disease; PUD, peptic ulcer disease; UTI, urinary tract infection.

Clinical Features

- Constitutional symptoms are condition-dependent and can take priority over pain issues in determining proper diagnostic studies and treatment options.

NONORGANIC SOURCES OF BACK PAIN

General

- Patients may exhibit exaggerated complaints with a nonanatomic basis and without an organic pathology. Multiple screening tests exist. In particular for patients with low back pain are the Waddell's signs.
- Waddell's signs are designed to delineate a nonorganic component for the patient's low back pain:
  - Demonstration of more than three out of five signs may be cause for suspicion.
  - These signs can be remembered with the acronym DO ReST.
  - Be aware that an organic component is not excluded with positive Waddell's signs.
  - It also does not diagnose any specific disorders.

🕮 WADDELL'S SIGNS

(DO ReST)	COMMENTS
Distraction	Presentation of severe radicular pain with the supine straight leg raising test but no pain in the seated straight leg raising test. Both should be positive.
Overreaction	Inappropriate, disproportionate reactions to a request. This may manifest with exaggerated verbalizations, facial expressions, tremors, or collapsing.
Regionalization	Motor or sensory abnormalities without anatomic basis, such as in a stocking-glove distribution, give-way to weakness, or a cog-wheel type of rigidity.
Simulation	Leg or lumbar pain with a light axial load on the skull. Or a presentation of lumbar pain with simultaneous pelvis and shoulder rotation in unison.
Tenderness	Exaggerated sensitivity or dramatic reproduction of pain with light touch of the soft tissue or with skin rolling.

MALINGERING

General

- Patients may misrepresent their condition due to secondary gain issues. More than pure symptom magnification or a deceptive distortion of events, malingering is a *Diagnostic and Statistical Manual of Mental Disorders* (5th ed.; *DSM-5*; American Psychiatric Association, 2013) disorder.
- Malingering is defined as an intentional production of falsely or grossly exaggerated physical and psychological symptoms for primary or secondary gain.
- Criteria for diagnosing malingering are defined by the *DSM-5*.

Etiology

- Motivated by external incentives.
- Avoiding work.
- Avoiding military duty.
- Obtaining financial compensation.
- Obtaining drugs.
- Evading criminal prosecution.

Imaging

- There are no specific studies to determine if a patient is malingering or demonstrating associated disorders. Certain psychological tests may offer insight on a patient's condition, but the diagnosis rests mainly on clinical suspicion and the exclusion of organic etiologies.

Treatment

- This rests on addressing the underlying issues involved with each patient's individual situation. It may require a multidisciplinary approach incorporating diverse aspects of the medical field, as well as confronting certain social matters.

INTERVENTIONAL SPINAL PROCEDURES

- 🕮 Percutaneous diagnostic and therapeutic spinal techniques continue to evolve in the management of spinal pain. Knowing the benefits and consequences of these procedures is an important aspect of delivering high-quality patient care, improving prognosis, and enhancing quality of life. This section serves as an introduction to these procedures, which can be considered in the comprehensive approach to spinal rehabilitation.
- Please read Interventional Spinal Procedures section in Chapter 11, Pain Medicine, for further discussion.

PATIENT SELECTION

- A complete patient history and physical exam, supported by appropriate diagnostic studies such as advanced spinal imaging and electrodiagnostic studies, must provide evidence to confirm the physician's choice of management.
- Essential prior to performing any interventional procedure is the screening for any serious occult pathology such as active infections, uncontrolled diabetes, tumors, and other disease processes.

🕮 COMPLICATIONS

- A thorough understanding of the procedural risks is paramount in providing appropriate care. It is outside the scope of this section to review all associated problematic outcomes.
- Confronting issues associated with vasovagal episodes, anaphylactic/allergic reactions, various infections, epidural hematomas, dural puncture headaches, spinal blocks, pneumothorax, respiratory depression, seizures, cerebral/cerebellar/spinal cord infarction or compression, causing paralysis, and death must be anticipated.
- Therefore, understanding the complications associated with intravascular, intrathecal, subdural, intraneural, intraosseous, and intrapleural compromise must be appreciated.
- Minor complications (Botwin et al., 2003):
  - Increased axial pain
  - Nonpositional headache
  - Facial flushing
  - Vasovagal (which may require cessation of the procedure and supportive care with Trendelenburg position, and increase IV fluids)
  - Superficial skin infection
  - Insomnia
  - Nausea and vomiting
- Major complications (Botwin et al., 2003):
  - Epidural hematoma
  - Cushing's syndrome
  - Epidural lipomatosis—can be self-limited
  - Subdural block
  - Intrathecal block
  - Seizure
  - Direct needle trauma
  - Spinal cord infarction

- Stroke
- Blindness
- Death

DIAGNOSTIC INTERVENTIONAL PROCEDURES

Diagnostic interventional procedures can help identify or rule out a particular structure as a spinal pain generator.

Diagnostic Medial Branch Blocks (Figure 4–196)

- A purely diagnostic test that evaluates the facet joint as the potential pain generator by anesthetizing the medial branches of the dorsal rami innervating that particular joint. Pre- and postprocedure pain scores should be taken.
- It primarily serves to formally diagnose facet syndrome. It is a prerequisite prior to performing an RFA.
- It is *not* considered a therapeutic intervention.

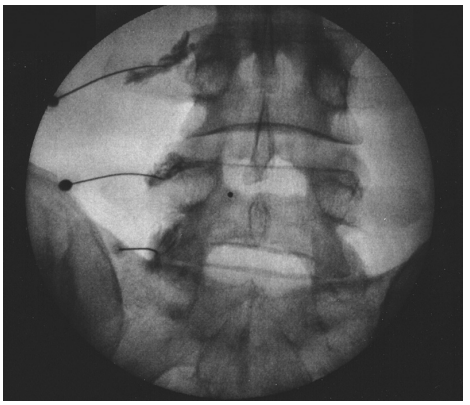

FIGURE 4–196 Lumbar medial branch block.

Provocative Discography (Figure 4–197)

- Diagnostic procedure to establish or rule out the intervertebral disc as a primary pain generator of axial spinal pain with or without radicular symptoms.
- Due to the significant rate of false positives, its diagnostic utility remains controversial. Furthermore, studies have shown accelerated disc degeneration in patients who have undergone discography testing.
- The procedure is performed by injecting contrast material into the nucleus pulposus of a disc; the disc is pressurized with contrast injectate:
  - Concordant pain is produced in the abnormal disc due to intolerance of increased intradiscal pressures or contrast material leaking through annular fissures reaching nociceptor fibers. Note that nonconcordant pain may be elicited and should not be considered a positive result.
  - Grading is based on Dallas or Modified Dallas Discogram Classification.

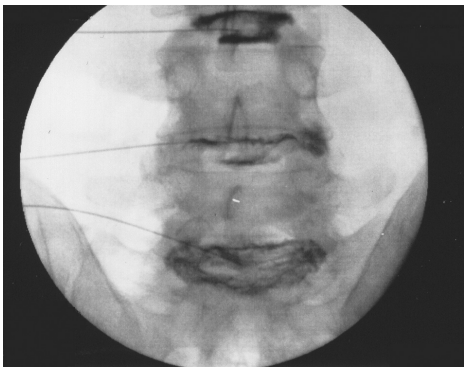

FIGURE 4–197 Provocative lumbar discography.

Selective Spinal Nerve Root Blocks (Figure 4–198)

- Diagnostic test that anesthetizes a specific spinal nerve to confirm or rule out a particular spinal nerve root as the primary pain generator.
- It can be used if the patient presents with radicular symptoms, but diagnostic studies (e.g., MRI, EMG) do not show specific, corroborating findings.
- It can be used if the patient presents with multilevel pathologies that interfere with distinguishing an accurate diagnosis, that is, generalized spinal stenosis, or multiple disc herniations.

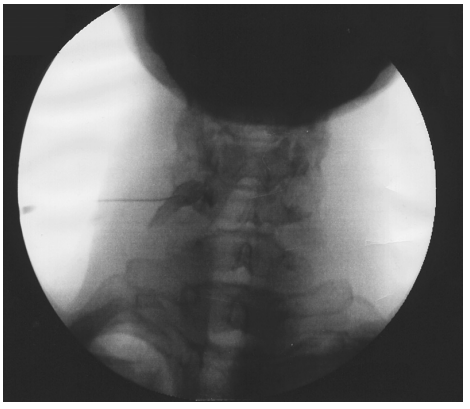

FIGURE 4–198 Cervical spinal nerve block.

Diagnostic Sacroiliac Joint Blocks

- Diagnosing pain from the SI joint region can be difficult. There is no widely accepted gold standard from a clinical and imaging standpoint. Significant disparities exist in studies examining the reliability, sensitivity, and specificity of clinical exam maneuvers.
- To diagnose SI joint pain, The Interventional Pain and Spine Intervention Society guidelines recommend placebo-controlled diagnostic intra-articular blocks to help confirm or rule out the SI joint as the primary pain generator. This is performed by an intra-articular SI joint block with high-dose anesthetic to see whether this joint is the source of pain. It can be used to help distinguish it from facet or discogenic-mediated spinal pain.

Sympathetic Blocks (Figure 4–199)

- Sympathetic blocks primarily serve to help establish a diagnosis of sympathetic-mediated pain syndromes (e.g., complex regional pain syndrome [CRPS]) by anesthetizing specific sympathetic ganglia.
- Sympathetic blocks test if the upper/lower extremity or trunk pain is sympathetically mediated. This is performed by anesthetizing the sympathetic fibers at specific locations anterior to the vertebral bodies.
- It primarily serves to establish a diagnosis of sympathetic-mediated pain syndromes (e.g., CRPS) by resetting the normal sympathetic tone.
- It may have longer lasting therapeutic effects, as well as serve as a precursor to performing an RFA.

THERAPEUTIC INTERVENTIONAL PROCEDURES

- Therapeutic interventional spinal procedures can offer the patient long-term relief by reducing inflammation or control a chronic pain condition. These treatments are typically part of a comprehensive program focused on optimizing functional restoration. They are typically

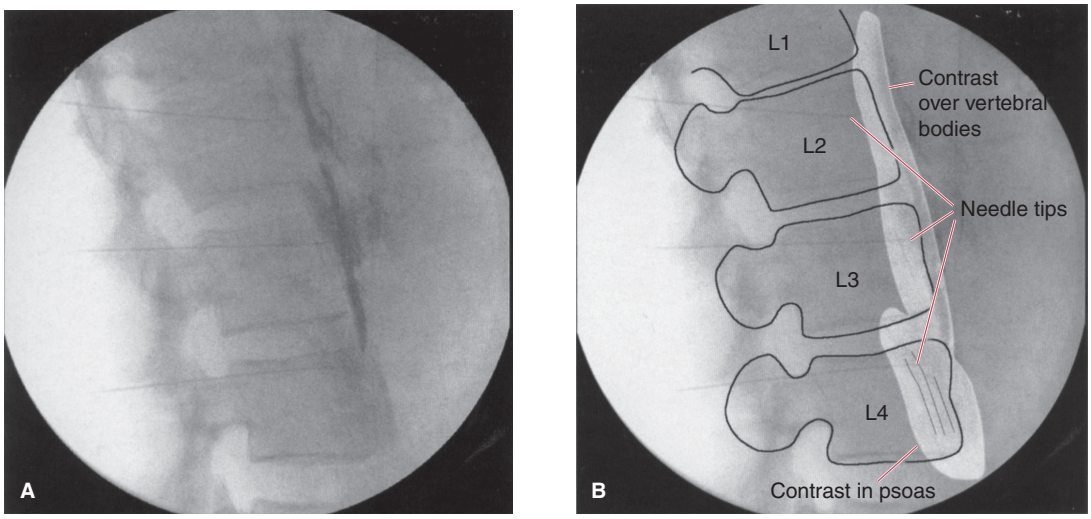

FIGURE 4–199 Lateral fluoroscopy view during neurolytic lumbar sympathetic block. (A) Three needles are in position with their tips over the anterolateral surfaces of L2, L3, and L4. 1 mL of contrast dye has been placed through each needle. Contrast has spread tightly adjacent to the anterolateral surface of the vertebral bodies through the needles at L2 and L3. The contrast adjacent to the needle at L4 has spread more diffusely in an anterior and inferior direction, indicating injection within the psoas muscle. This needle must be repositioned before neurolysis in a more anterior and medial direction. Neurolysis is carried out by placing 2 to 3 mL of neurolytic solution (10% phenol in iohexol 180 mg/mL or 50%–100% ethyl alcohol) through each needle. The needle position for RF neurolysis is identical. (B) Labeled image.
RF, radiofrequency.
Source: From Rathmell JP. *Atlas of Image Guided Intervention in Regional Anesthesia and Pain Medicine*. Lippincott Williams & Wilkins; 2006, with permission.

paired with appropriate pharmacotherapeutics, physical therapies, and screening for psychological disorders.

Facet (Zygapophyseal) Joint Injections (Figure 4–200)

- The diagnosis of facet-mediated pain has been problematic, as clinical examination findings or spinal imaging does not reliably diagnose facet-mediated pain.
- Confirmatory diagnostic blocks should be performed prior to consideration of therapeutic facet joint injections.
- The goal is to provide long-term pain relief from a particular facet joint by injecting corticosteroids intra-articularly after diagnostic blocks. This helps confirm a facet-mediated pain diagnosis.
- Therapeutic facet injections serve to inhibit inflammatory mediators within the facet joint, which may have been provoked by abnormal biomechanics, degenerative joint disease, trauma, and postlaminectomy syndrome.
- More recent studies have noted limitations with such an approach.

Facet Joint Medial Branch Radiofrequency Ablation (Figure 4–201)

- The goal is to provide longer term pain relief from facet-mediated spinal pain by ablating the medial branch nerves that innervate the facet.

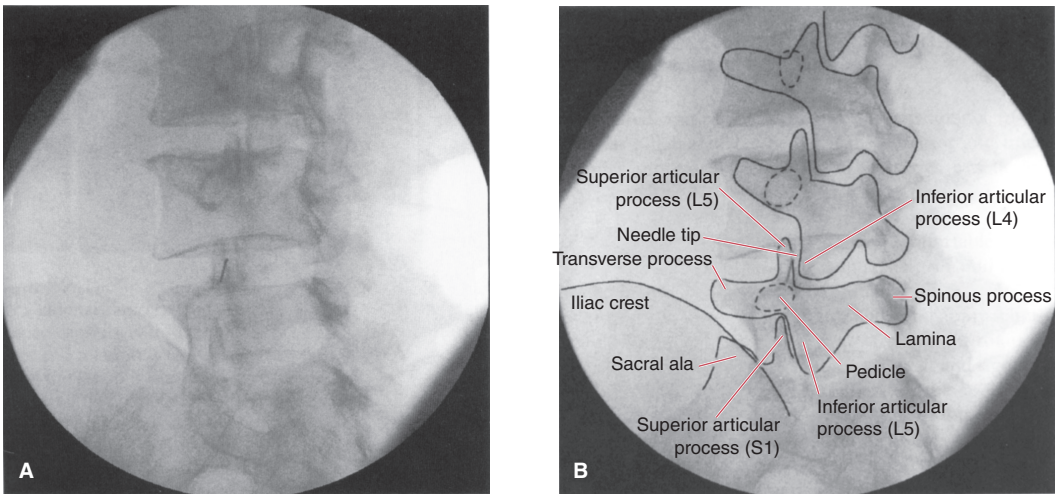

FIGURE 4–200 Oblique radiograph of the lumbar spine during lumbar intra-articular facet injection. (A) The needle is in place in the left L4–L5 facet joint. The needle travels from inferior to slightly superior. (B) Labeled image. *Source:* From Rathmell JP. *Atlas of Image Guided Intervention in Regional Anesthesia and Pain Medicine*. Lippincott Williams & Wilkins; 2006, with permission.

- Two positive diagnostic medial branch blocks are recommended to confirm the diagnosis.
- The radiofrequency (RF) probes are placed parallel to the medial branches that innervate the symptomatic facet joint. Denervation of the specific facet joint is achieved by thermocoagulation of the innervating medial branches.

Epidural Steroid Injections

- The goal is to provide long-term pain relief from radicular pain by treating the inflammatory component of a radiculopathy. This is performed by placing a corticosteroid solution around the affected spinal nerve root in the epidural space. It medicates the inflamed neural structures affected by pathologies such as disc herniations or spinal stenosis.
- **Transforaminal approach (Figure 4–202)**:
  This injection delivers a maximal concentration of medication to the target point via the neuroforamen. Landmarks have been described as needle placement in the ventral aspect of the

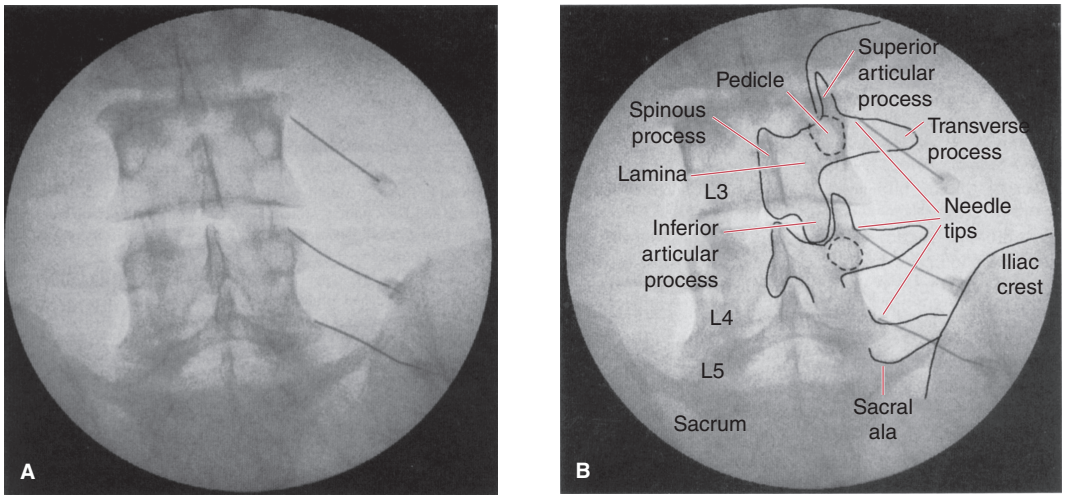

FIGURE 4–201 AP view of the lumbar spine during lumbar RF treatment of the lumbar facet joints. (A) Three RF cannulae are in place at the base of the TP and SAP at the L3, L4, and L5 vertebral body levels on the right. Note the angle of the cannulae is parallel to the medial branch nerve. (B) Labeled image.
AP, anterior-posterior; RF, radiofrequency; SAP, superior articular processes; TP, transverse processes.
Source: From Rathmell JP. *Atlas of Image Guided Intervention in Regional Anesthesia and Pain Medicine*. Lippincott Williams & Wilkins; 2006, with permission.

foramen in the thoracolumbar spine and dorsal placement in the cervical spine, to avoid the vertebral artery.

- **Interlaminar approach (Figure 4–203)**: This injection delivers the medication to the general epidural space at a particular level via a loss-of-resistance or hanging drop technique utilized through the interlaminar space. Landmarks consist of an epidural space bounded by the dura anteriorly and ligamentum flavum posteriorly. Loss of resistance in the syringe occurs when the ligamentum flavum is pierced, which is the site of injection. Epidural catheters can be used to assist in a more targeted placement of injectate.
- **Caudal approach (Figure 4–204):** This injection delivers the medication into the sacral canal and superiorly at most to the L4–L5 level. The entry point is between the sacral cornua through the sacrococcygeal ligament into the sacral hiatus. The needle is advanced to the level of the S3 neuroforamen to avoid the dura. Epidural catheters can be used to assist in a more targeted placement of injectate.

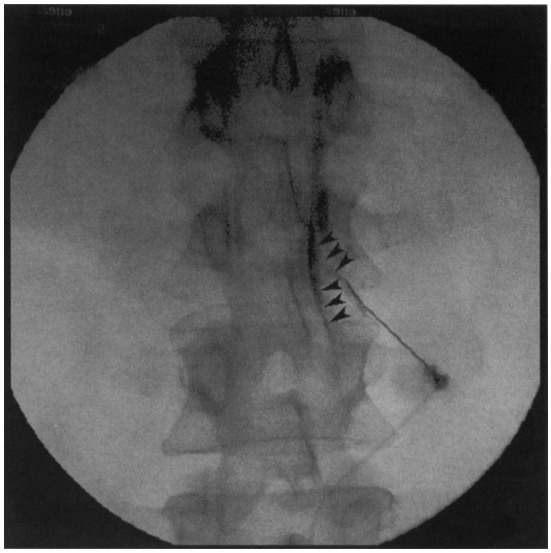

FIGURE 4–202 Lumbar transforaminal injection (AP view). The needle is in final position for right L3–L4 transforaminal injection following injection of contrast dye. The needle tip lies inferior to the pedicle, and contrast dye extends to the right lateral epidural space beneath the pedicle (upper group of arrowheads). Contrast also extends along the left lateral aspect of the epidural space to outline the right L4 nerve root as it exits through the lateral recess at L4–L5 (lower group of arrowheads).
AP, anterior-posterior.
Source: From Rathmell JP. *Atlas of Image Guided Intervention in Regional Anesthesia and Pain Medicine*. Lippincott Williams & Wilkins; 2006, with permission.

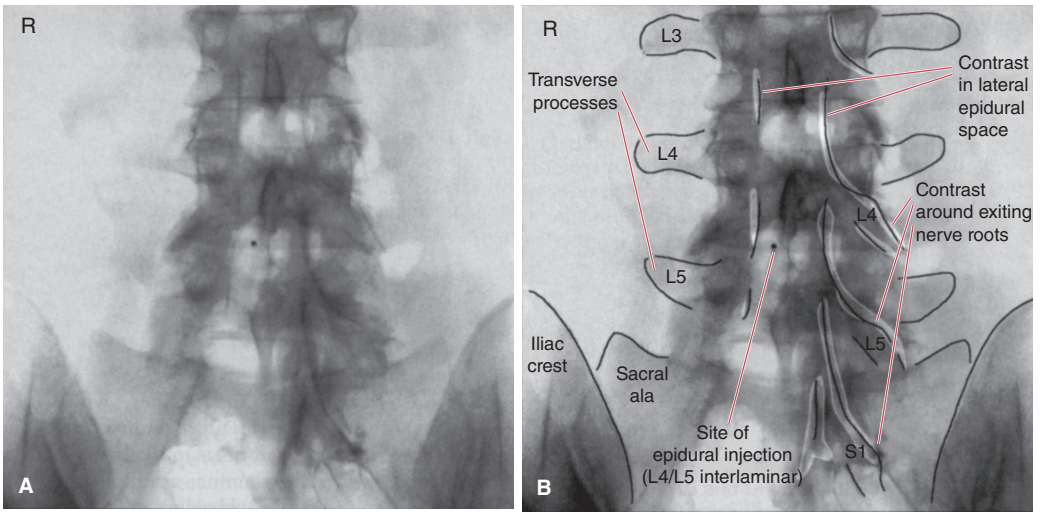

FIGURE 4–203 Lumbar interlaminar injection (AP view) epidurogram of the lumbosacral spine. (A) When larger volumes of injectate are used (in this image, 10 mL of contrast-containing solution), the injectate spreads extensively within the anterior and posterior epidural space and exits the intervertebral foramina, surrounding the exiting nerve roots. However, in the presence of significant obstruction to flow, as in this patient with a right L4/L5 disc herniation and compression of the exiting right L4 nerve root, the injectate often follows the path of least resistance, exiting the foramina on the side opposite from the disc herniation. (B) Labeled image.
AP, anterior-posterior.
Source: From Rathmell JP. *Atlas of Image Guided Intervention in Regional Anesthesia and Pain Medicine*. Lippincott Williams & Wilkins; 2006, with permission.

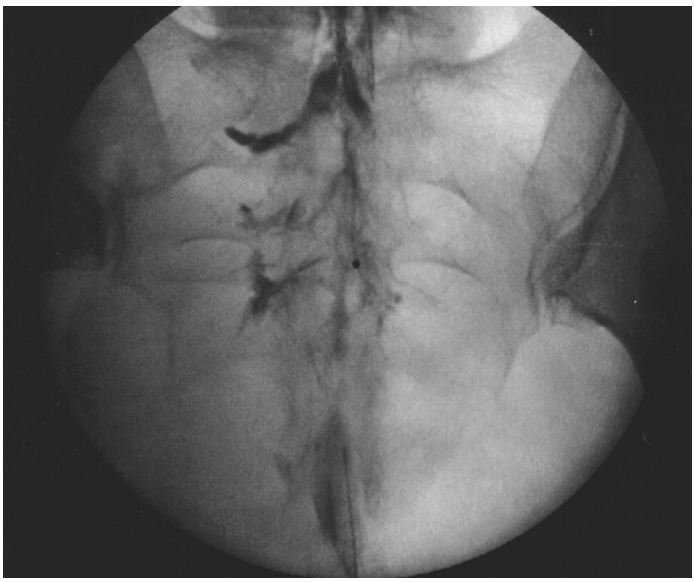

FIGURE 4–204 Caudal epidural injection.

Therapeutic Sacroiliac Joint Injections (Figure 4–205)

- Goal is to provide pain relief for the SI joint via intra-articular corticosteroid injection.
- It serves to inhibit inflammatory mediators within the joint, which may have been provoked by abnormal biomechanics (e.g., degenerative joint disease, trauma, postlumbar fusion) or spondyloarthropathies.

Sacroiliac Joint Radiofrequency Neurotomy

- Goal is to provide longer pain relief for the SI joint. This is performed by placing a Teflon-coated electrode on a specific area for the L5–S3 lateral branches of the dorsal sacral plexus.

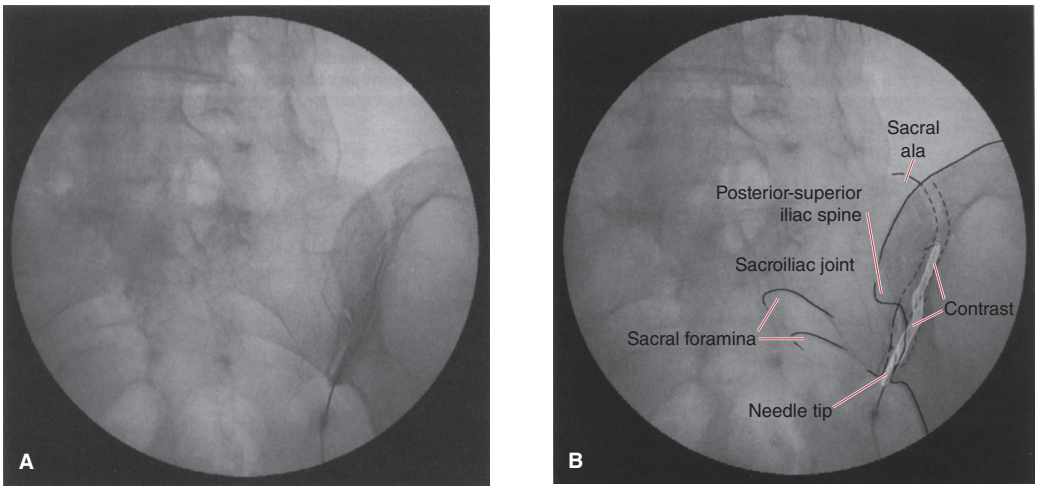

FIGURE 4–205 AP view of a right intra-articular SI joint injection. (A) A 22-gauge spinal needle is in position in the posterior inferior aspect of the right SI joint, and 1.5 mL of contrast dye has been injected. Contrast extends to the superior portion of the joint. (B) Labeled image.
SI, sacroiliac.
Source: From Rathmell JP. *Atlas of Image Guided Intervention in Regional Anesthesia and Pain Medicine*. Lippincott Williams & Wilkins; 2006, with permission.

- It prohibits the SI joint from sensing pain by coagulating its neuronal innervations.
- Evidence of efficacy of SI joint RF neurotomy is limited due to lack of prospective randomized controlled trials and lack of standardized lesion techniques (Aydin et al., 2010).

Intradiscal Treatments

- The purpose of these procedures is to provide long-term pain relief of either discogenic or radicular pain. They are performed by placing a specialized device into the disc to alter the annular integrity or decrease intradiscal pressure.
- **IDET (Figure 4–206A)**:
  - This treatment predominantly focuses on discogenic sources of spinal pain. A blunt-tipped thermal catheter is threaded through an introducer cannula into the nucleus, traversing the posterior aspect of the disc at the nuclear–annular junction. Thermocoagulation across annular fissures has been proposed to ablate disc nociceptors, remodel collagen fibers, and denature inflammatory mediators.
  - The current literature does not show significant efficacy of IDET over placebo.
- **Percutaneous disc decompression (Figure 4–206B)**: This treatment focuses on discogenic and radicular pain. A specialized device is threaded through an introducer cannula into the nucleus pulposus. The removal of this material has been proposed to reduce intradiscal pressure, to unweight internal disc nociceptors or the spinal nerve roots (e.g., Nucleoplasty, Nucleotome, DeKompressor):
  - There is a paucity of high-quality literature for percutaneous disc decompression, notably a lack of blinded, randomized studies (Manchikanti et al., 2009).

Implantable Therapies

- The goal of these devices is to provide prolonged pain relief by modulating pain signal transmission or with continuous delivery of analgesic medication.
- 🕮 **Spinal cord stimulator (SCS; Figure 4–207)**: Modulates pain signals in the spinal cord primarily through the gate control theory:
  - SCS implantation should be considered only after an SCS trial has been performed with successful results.
  - After a successful SCS trial, a permanent SCS may be implanted. The SCS electrodes are placed over the area of the dorsal columns into the epidural space.

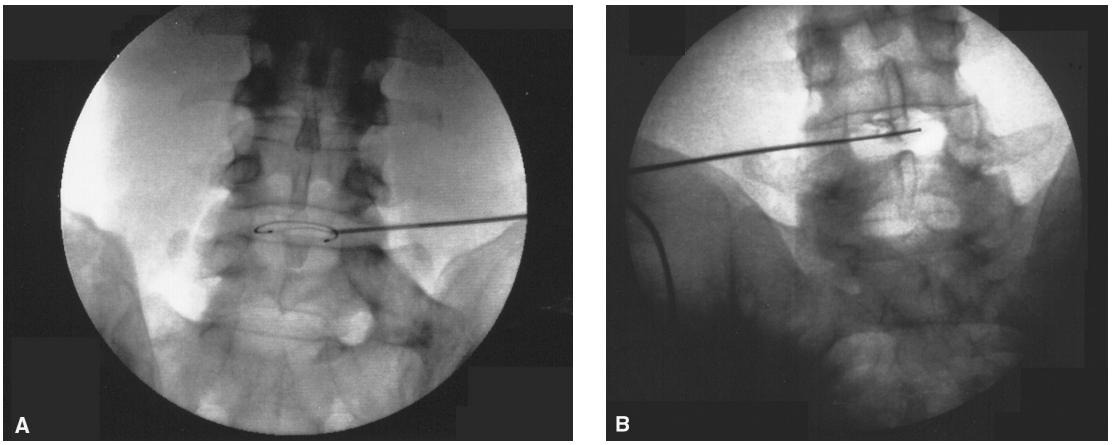

FIGURE 4–206 Intradiscal treatments. (A) Lumbar IDET. (B) Percutaneous disc decompression (lumbar nucleoplasty). IDET, intradiscal electrothermal therapy.

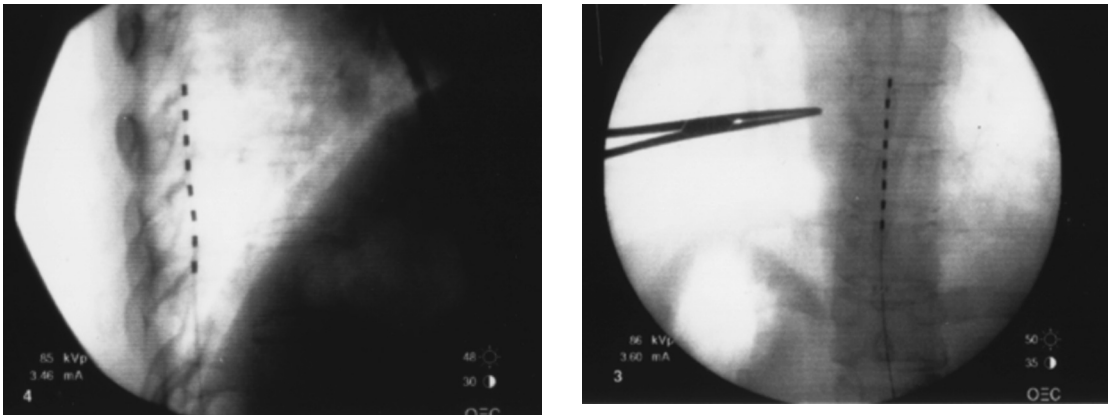

FIGURE 4–207 AP and lateral view of a SCS.
Ap, Anterior-Posterior; Scs, Spinal Cord Stimulator.

- **Dorsal root ganglion (DRG) stimulator:** Modulates pain signals at the DRG through an altered mechanism of the gate control theory, being numerous nerve fibers can be stimulated simultaneously (Figure 4–208).

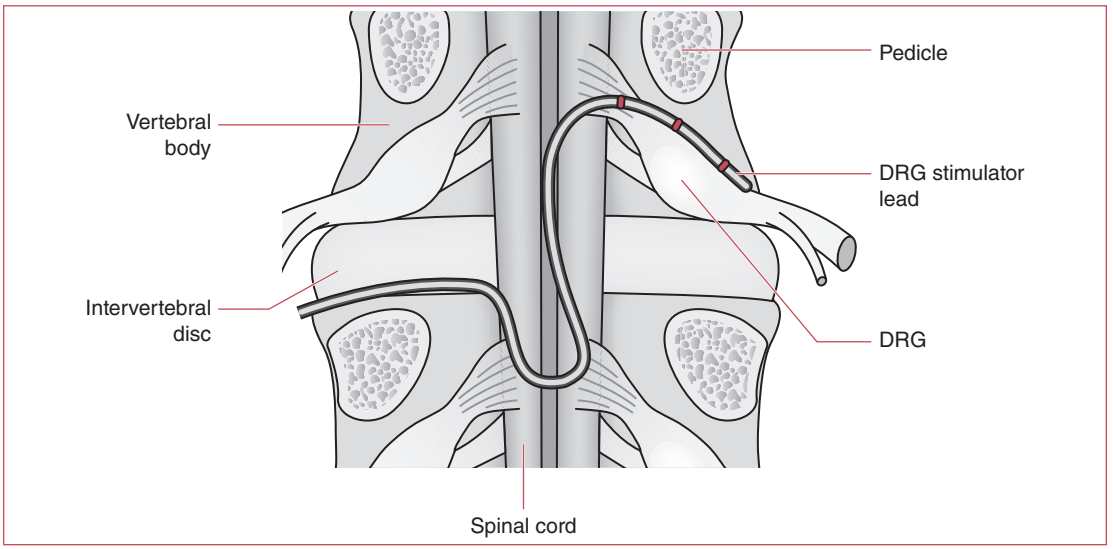

FIGURE 4–208 DRG stimulator.
DRG, dorsal root ganglion.

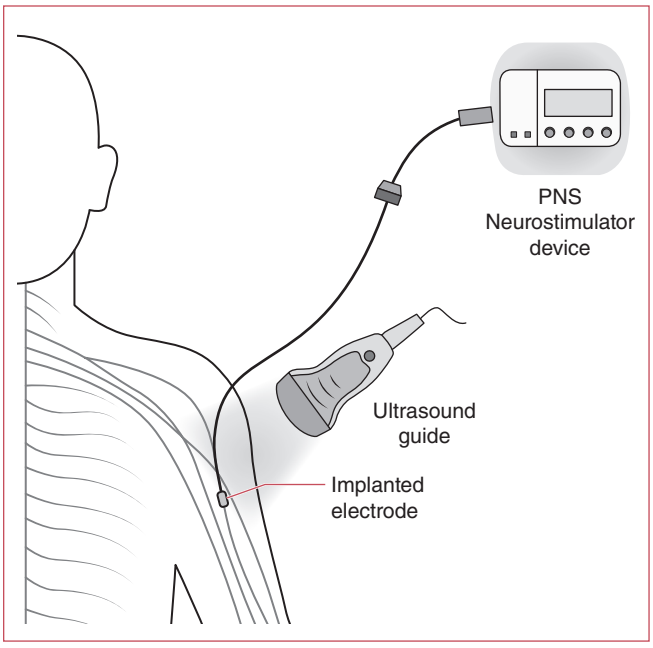

FIGURE 4–209 Peripheral nerve stimulator (PNS).

  - DRG stimulation is a new neuromodulation modality that provides selective neuromodulation of the DRG and is FDA-approved to treat CRPS.
  - It can localize treatment to a specific dermatome in treating neuropathic pain syndromes.
    - Neuropathic pain populations may include post hernia repair, total knee replacement, amputee pain, pelvic pain, and foot pain.
  - After a successful DRG stimulator trial, a permanent stimulator device can be implanted.
  - However, the current literature is limited and dependent on industry evaluation (Potter et al., 2022).
- **Peripheral nerve stimulator (PNS):** Neuromodulation systems that incorporate peripheral and central mechanisms to create and analgesic effect (**Figure 4–209**).
  - PNS targets specific peripheral nerves placed under image guidance (US or fluoroscopy).
  - The device consists of a permanent or temporary percutaneously placed electrode and external pulse generator.
  - Utilized in peripheral nerve injuries or joint pains axial spinal pain and headaches, improved by a peripheral nerve block or stimulation test.
  - However, there is a paucity of high-quality literature supporting its use and limited data on long-term efficacy and risks.
- **Intrathecal pain pump:** Medications such as opioids (morphine or hydromorphone, sufentanil, fentanyl, methadone), local anesthetics (bupivacaine, ropivacaine), and alpha-2 adrenergic agonists (clonidine) are delivered directly to spinal receptors to mediate pain (**Figure 4–210**):
  - This is performed by placing a specialized catheter into the intrathecal space, which is connected to a pump/reservoir system in the subcutaneous tissue of the abdomen.

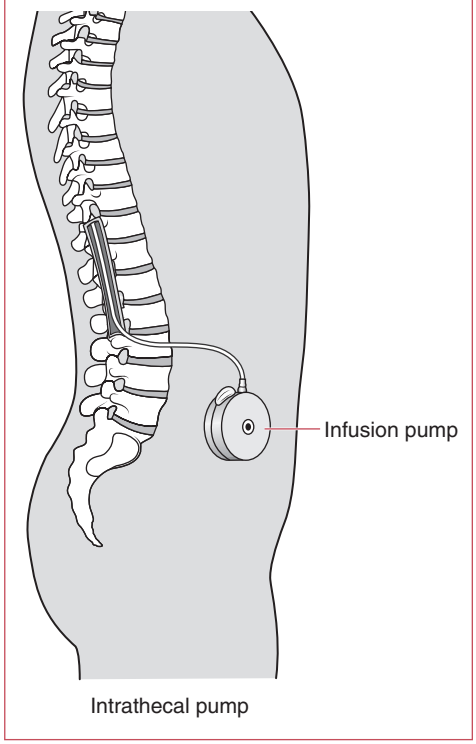

FIGURE 4–210 Intrathecal pain pump.

Epidural Lysis of Adhesions (Epidural Adhesiolysis)

- This procedure is indicated for spinal pain with or without radicular pain due to adhesions corroborated by diagnostic imaging (Figure 4–211).
- This is performed by placing a semirigid catheter into the epidural space via a caudal, interlaminar, or transforaminal approach. It delivers medications to the region of the adhesions to decrease inflammation and enhances breakdown of epidural fibrosis.
- Contraindications include infection, coagulopathy, and presence of arachnoiditis.
- There is a paucity of literature and limited evidence showing efficacy in treating postsurgery syndrome.

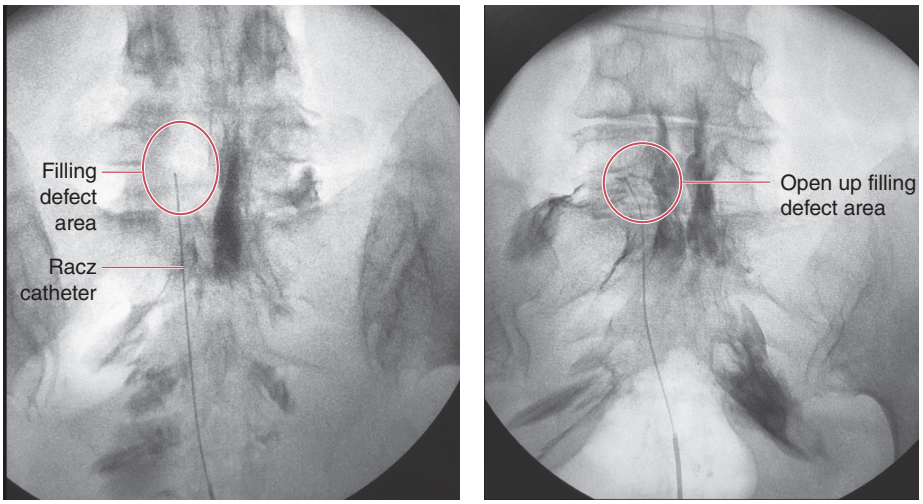

FIGURE 4–211 Epidural lysis of adhesions.
Source: From Ege F. Evaluation of the efficacy of caudal epidural neuroplasty in patients with lumbar epidural fibrosis. *Cureus*. 2024;16(1):e52606. doi:10.7759/cureus.52606. PMID: 38260106; PMCID: PMC10803106.

Minimally Invasive Lumbar Decompression

- Focused on patients with lumbar spinal stenosis (LSS) caused by ligamentum flavum hypertrophy.
- A procedure performed under fluoroscopic guidance to posteriorly gain access of a ligamentum flavum and debulk the tissue using percutaneous decompressive devices (Figure 4–212).
- The procedure is aimed at relieving neurogenic claudication symptoms in the lower extremities associated LSS.
- While there are limited studies showing efficacy, there are no high-quality studies demonstrating long-term benefits (Diwan et al., 2019).

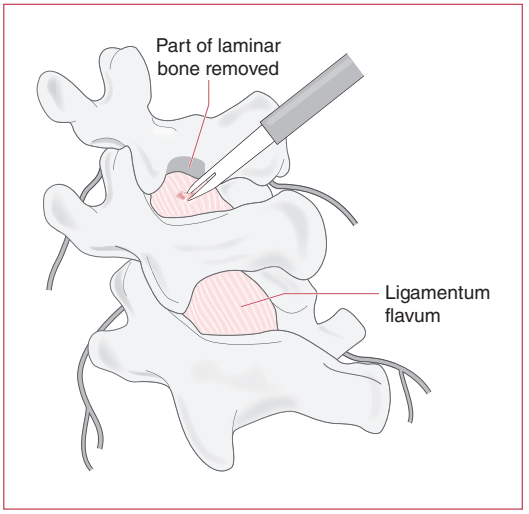

FIGURE 4–212 Minimally invasive lumbar decompression.

Vertebral Augmentation

- Goal is to provide pain relief from vertebral body compression fractures, for people who have minimal to no pain relief from 4 to 6 weeks of conservative treatments. The most common etiology of a fracture is osteoporosis. Other causes described are metastatic disease, multiple myeloma, expanding hemangiomas, Paget's disease, and painful (acute) Schmorl's nodes.
- This procedure is performed by injecting polymethyl methacrylate (PMMA) cement into the fracture site:
  - **Vertebroplasty**: PMMA is injected directly into the vertebral body through an introducer cannula. Its primary focus is pain relief.

INDICATIONS AND CONTRAINDICATIONS FOR VERTEBROPLASTY

INDICATION	CONTRAINDICATION
Painful osteoporotic compression fracture Osteolytic metastasis Multiple myeloma Symptomatic hemangioma	Cord compression with clinical myelopathy Active infection Bleeding disorder (coagulopathy) Asymptomatic fracture

  - **Kyphoplasty (Figure 4–213)**: A balloon tamp/introducer cannula is placed, and PMMA is indirectly injected into the vertebral body through the balloon. Its goal is pain relief and restoration of vertebral body height.

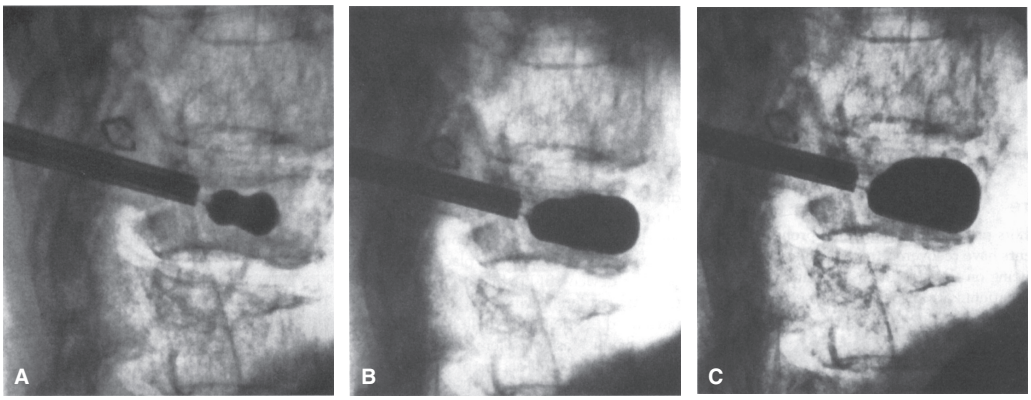

FIGURE 4–213 Kyphoplasty procedure. Inflation continues (A–C) until vertebral body height is restored. The inflatable bone tamp (IBT) contacts a vertebral body cortical wall, and the IBT reaches maximal pressure rating without spontaneous decay, or the maximal balloon volume is reached.
Source: From Slipman CW, Derby R, Simeone F, et al. *Interventional Spine: An Algorithmic Approach.* Saunders; 2008, with permission.

- For osteoporotic fractures, percutaneous vertebral augmentation has been increasingly utilized, but the long-term efficacy, cost-effectiveness, and safety of vertebroplasty and kyphoplasty remain unclear based on the current literature. More recent, randomized controlled studies comparing vertebroplasty with sham showed no significant difference in outcomes for patients with osteoporotic vertebral body fractures (Buchbinder et al., 2009; Kallmes et al., 2009; Wardlaw et al., 2009). Additional studies are warranted to clarify its utility in treating osteoporotic fractures.

Basivertebral Nerve Radiofrequency Ablation

- The basivertebral nerve is a branch of the sinuvertebral nerve that enters the vertebral body and innervates the endplates.

- Painful degenerative vertebral endplates may be seen in Modic Type I and Type II changes seen on T2-weighted images on MRI, may correlate with vertebrogenic pain.
  - It is important to note that Modic changes are routinely seen on MRI without symptoms, and the significance of Modic changes continues to be debated (Rahme & Moussa, 2008).
  - There is no gold standard to diagnose vertebral endplate pain.
- Interosseous RFA of the basivertebral nerve to denervate targeted nociceptive fibers within the vertebral body causing anterior column pain at the L3–S1 levels (**Figure 4–214**).
- Studies show potential short-term benefit, but there are no controlled, high-quality, long-term studies without industry involvement that demonstrate efficacy.

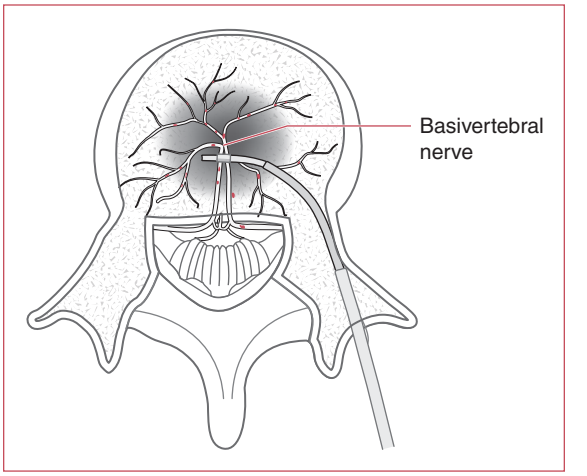

FIGURE 4–214 Basivertebral nerve RFA. RFA, radiofrequency ablation.

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ELECTRODIAGNOSTIC MEDICINE AND CLINICAL NEUROMUSCULAR PHYSIOLOGY

5

Ted L. Freeman, DO • Ernest W. Johnson, MD[1] • Eric D. Freeman, DO • David P. Brown, DO • Lei Lin, MD, PhD

INTRODUCTION

Electrodiagnostic (EDX) medicine should be considered an extension of a comprehensive patient history and physical examination. Combining data found on nerve conduction studies (NCS) and needle electromyography (EMG), the pathophysiology of a peripheral nerve disease process can be further defined to illustrate location, duration, severity, and prognosis. It can function as a valuable aid in patient management, serving as an extension of the clinical exam, but not a substitute.

This chapter focuses on board-related topics about EDX medicine as well as neuromuscular disorders and their associated electrophysiologic changes. It is to be used as a study guide and is not intended to be an all-inclusive composite. For more elaborate coverage of the subject matter, the reader is directed to the References and Recommended Reading sections at the end of this chapter.

BASIC PERIPHERAL NERVOUS SYSTEM ANATOMY

NEURON ANATOMY AND FUNCTION

- **Cell body:**
  - The cell body (or soma) of a motor or sensory nerve
  - Cell bodies of motor neurons are located in the anterior (ventral) horn region of the spinal cord and project an axon distally. It regulates the characteristics of the entire motor unit.
  - Cell bodies of sensory neurons are bipolar cells with two axons (one axon projects proximally and the other distally) and are found in the dorsal root ganglion (DRG), which is located outside the spinal cord in the proximity of the intervertebral foramen.
- **Axon:**
  - This is the projection from the sensory or motor nerve cell body that propagates current flow and transports cell nutrition (axonal transport). It can be unmyelinated or myelinated by Schwann cells.

[1]Deceased

- At each spinal level, axons from motor and sensory neurons form ventral and dorsal nerve roots, respectively, which then combine to become a mixed (sensorimotor) spinal nerve. Each spinal nerve then branches off to a dorsal and ventral ramus.
- Motor axons project distally from their cell bodies to become motor nerve roots.
- Sensory axons project proximally to the spinal cord and distally to become sensory nerve roots.
- Myelin sheaths that cover an axon are electrical insulators that help to accelerate electrical signal conduction along the axon.

- **Nerve:**
  - A nerve is a bundle of axons that transmit signal to and from various parts of the body. Sensory nerves transmit sensory signals from the body to the central nervous system (CNS). Motor nerves transmit motor signals from the CNS to the body's skeletal muscles.
  - Nerves are covered by nerve connective tissue.
- **Peripheral nerves:**
  - Motor and sensory nerve fibers combine at various levels in the body (spinal nerve, ventral ramus, plexus) and ultimately terminate as peripheral nerves.
  - A peripheral motor nerve consists of multiple neural branches from the distal portion of the axon. They innervate individual muscle fibers.
  - 🕮 The amount of muscle fibers belonging to an axon is the innervation ratio (IR). This ratio varies, depending on the function of the motor unit.
  - Muscles of gross movement have a larger amount of their fibers innervated by one axon (high ratio). Muscles of fine movement have a smaller amount of their fibers innervated by one axon (low ratio).
    - The axons innervating leg muscles can have a ratio of 600 muscle fibers to one axon (600:1), while the IR of the eye muscles can be 1 muscle fiber to 1 axon (1:1).
    - The higher the IR, the greater the force generated by that motor unit. A myotome is a group of muscles that are innervated by one spinal segment.
  - Sensory nerves innervate various segments in the body and are arranged into spinal segmental levels of innervation known as dermatomes.
- **Neuromuscular junction (NMJ):**
  - Motor nerves synapse with muscle fibers at sites known as NMJs.
  - These sites are where the electric impulse propagated along the axon is converted into a chemical reaction. The signal is then translated back into an electrical impulse at the postsynaptic membrane to initiate muscle fiber action potentials (APs).
- 🕮 **Muscle fibers:**
  - These extrafusal fibers are the final components of the motor unit (see later section, The Motor Unit). Here, the electrical signal from the postsynaptic NMJ membrane stimulates muscle fiber depolarization and muscle fiber APs.
  - Muscle fiber characteristics, including twitch response, depend upon the type of alpha motor neuron by which it is innervated.

Nerve Connective Tissue (Figure 5–1)

- **Endoneurium:**
  - This is the connective tissue surrounding each individual axon and its myelin sheath.
  - The main function of the endoneurium is to electrically isolate individual nerve axons. It also plays an important role in maintaining endoneurial fluid pressure.

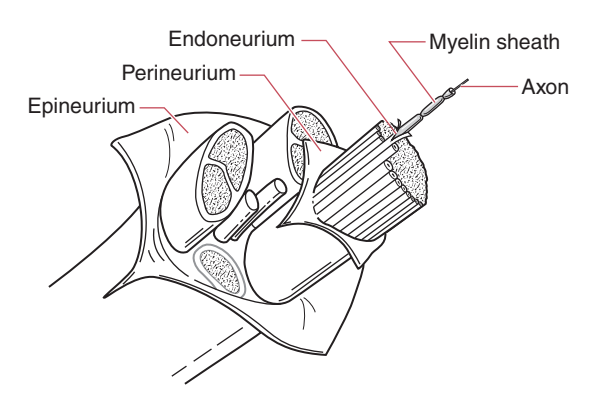


FIGURE 5–1 Neuronal connective tissue: The internal anatomy of the nerve.

- 🕮 It may help to keep the damaged ends of the axons in correct alignment and may facilitate repair of injured axons.

– **Perineurium:**
- This is the strong, protective, connective tissue surrounding bundles or fascicles of myelinated and unmyelinated nerve fibers.
- It helps strengthen the nerve and acts as a diffusion barrier. Individual axons may cross from one bundle to another along the course of the nerve.
- 🕮 If the perineurium is damaged, the contents bulge outward into the epineurium and the larger myelinated fibers become demyelinated in the underlying region.

– **Epineurium:**
- This is the loose connective tissue surrounding the entire nerve that holds the fascicles together and protects it from compression. This must remain intact in order to allow for possible nerve regrowth (Pedrosa et al., 2017).

The Motor Unit (Figure 5–2)

- The motor unit is the basic functional element of the neuromuscular system. It consists of the following components:
  – Anterior horn cell (motor nerve cell body)
  – Motor nerve axons
  – Peripheral nerve
  – NMJ
  – Muscle fibers

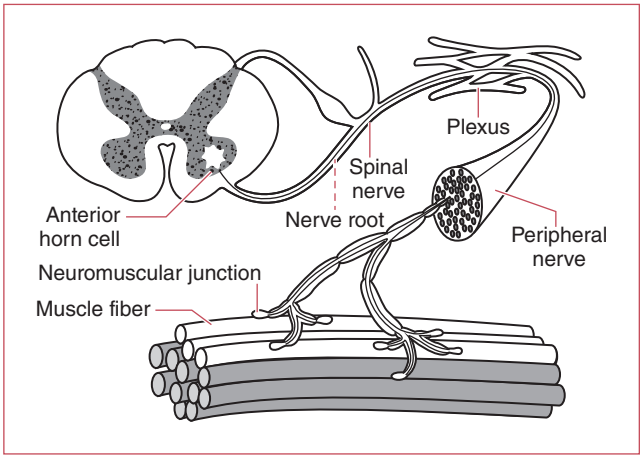


FIGURE 5–2 The motor unit.

Alpha Motor Neurons

- The three motor neurons listed in Table 5–1 innervate specific fibers, extrafusal or intrafusal.
- Needle EMG monitors factors related to the motor unit and thus is limited to evaluating the alpha motor neurons. The alpha motor neurons and associated motor unit parameters have been described based on size and physiology (Figure 5–3).
- The order of recruitment is related to their size, starting with the smaller motor units. This sequential activation allows for a smooth increase of contractile force and is described by the Henneman Size Principle.

Henneman Size Principle
- A smaller alpha motor neuron has a lower threshold of excitation, causing it to be recruited first during voluntary contraction.
- A larger alpha motor neuron has a higher threshold of excitation and is recruited when more motor units are needed to generate greater contractile force.

TABLE 5–1 Three Types of Motor Neurons

MOTOR NEURON	INNERVATIONS
Alpha	Extrafusal fibers—Skeletal muscle
Gamma	Intrafusal fibers—Muscle spindle
Beta	Intrafusal and extrafusal fibers

Muscle Fiber Types

FIBER TYPE	INNERVATION CHARACTERISTICS
Type I	Smaller cell body
	Thinner diameter axon
	Lower innervation ratio
	Slower twitch muscle fibers
Type II	Larger cell body
	Thicker diameter axon
	Higher innervation ratio
	Faster twitch muscle fibers

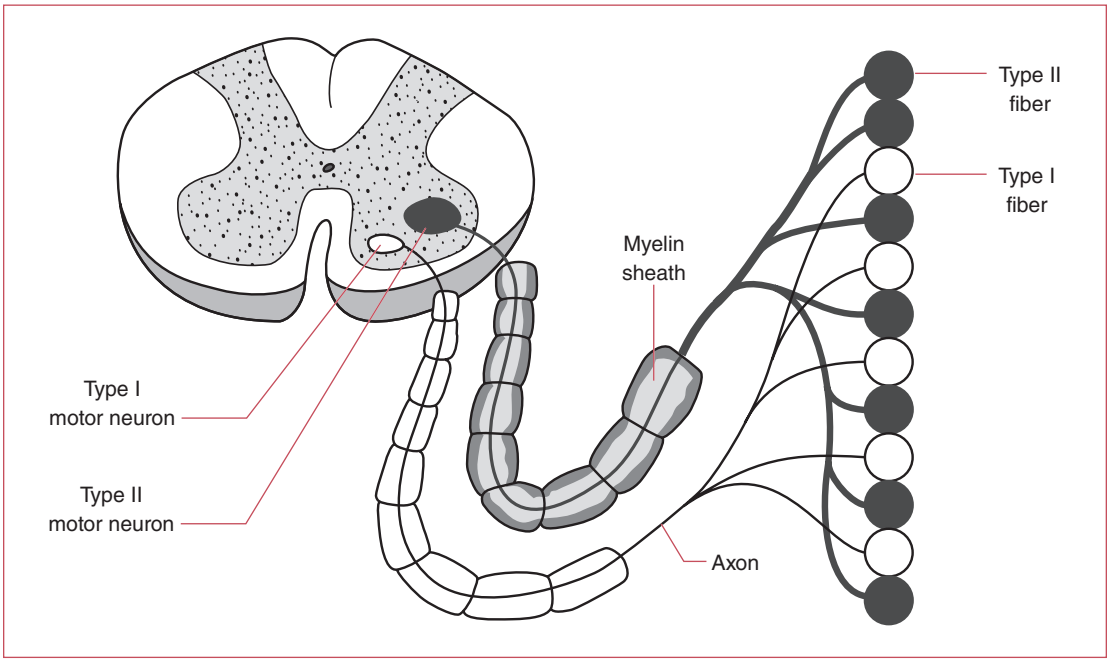


FIGURE 5–3 Description of Type I and Type II alpha motor neurons.

NERVE FIBER CLASSIFICATION (TABLE 5–2)

- Nerve fibers vary in their function based on their physiologic characteristics. Their classification is based on their diameter, conduction velocity (CV), and function.
- Table 5–2 describes two major classification systems that categorize the different nerve fibers.
- 🕮 EDX studies evaluate only Ia (large, myelinated) fibers.

TABLE 5–2 Nerve Fiber Classification

LLOYD AND HUNT (SENSORY)	ERLANGER AND GASSER (SENSORY AND MOTOR)	DIAMETER (μM)	VELOCITY (M/S)	FUNCTION
Ia fibers	A-alpha fibers	10–20 largest	50–120 fastest	Motor: Alpha motor neurons Sensory: Muscle spindle
Ib fibers	A-alpha fibers	10–20	50–120	Sensory: Golgi tendon organ, touch, pressure
II fibers	A-beta fibers	4–12	25–70	Motor: Intrafusal and extrafusal muscle fibers Sensory: Muscle spindle, touch, pressure
III fibers	A-gamma fibers A-delta fibers	2–8 1–5	10–50 3–30	Motor: Gamma motor neurons, muscle spindle Sensory: Touch, pain, temperature
IV fibers	B-fibers C-fibers	1–3 <1	3–15 <2	Motor: Postganglionic autonomic fibers Motor: Preganglionic autonomic fibers Sensory: Pain, temperature

NERVE PHYSIOLOGY

Resting Membrane Potential

- This is the voltage of the axon's cell membrane at rest. Normal resting membrane potential (RMP) is –70 to –90 mV.
- *Leak channels*:
  - These are openings in the cell membrane that allow sodium (Na^+) and potassium (K^+) to move passively in and out of the cell membrane.
- *Na^+–K^+ ATP-dependent pumps* (**Figure 5–4**):
  - A negative potential is maintained inside the cell by actively exporting three Na^+ ions while importing two K^+ ions through Na^+–K^+ ATP-dependent pumps located within the cell's semipermeable membrane.
  - This keeps each ion against a concentration gradient with a deficit of positive ions inside the cell. The RMP of the nerve would otherwise dissipate from the ions diffusing through the ion leak channels.

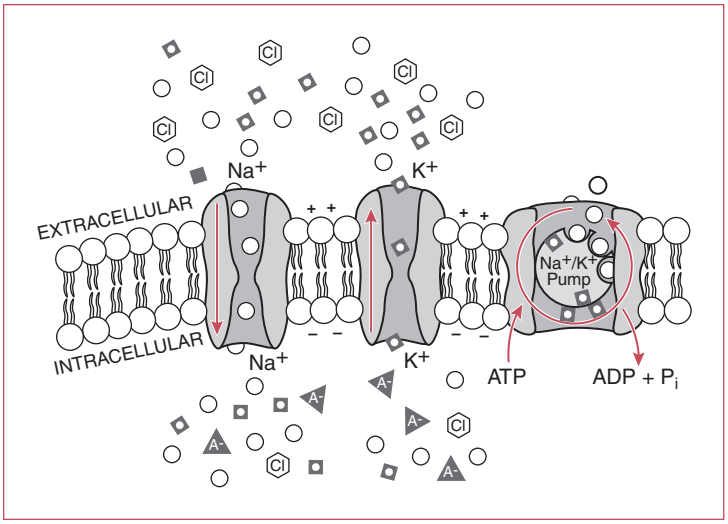


FIGURE 5–4 Na^+–K^+ ATP–dependent pump: (Two) K+ ions are imported. (Three) Na+ ions are exported; therefore, a negative potential is maintained inside the cell.
ADP, adenosine diphosphate; ATP, adenosine triphosphate.

Depolarization

- When an outside current is applied to a nerve by a stimulator consisting of a cathode (negative pole) and an anode (positive pole), positive charges on the axon become attracted under the cathode and lower the membrane potential.
- The membrane becomes increasingly permeable to Na^+, which rushes into the cell through the opened voltage-gated channels toward an equilibrium. This process of sodium conductance is the most important event in generating an AP.
- Action potential (**Figure 5–5**):
  - This is a voltage change occurring from an excited cell. The electric impulse propagates along an axon or muscle membrane. It can also be evoked by a stimulator. The all-or-none response travels in both directions along the axon.
- **All-or-none response:**
  - A stimulus must be strong enough to reach a certain threshold of activation. Once reached, the AP generated remains at a constant size and configuration.
  - If it is below this threshold, no potential will occur. Any stimulus intensity greater than the threshold will not generate a larger potential.

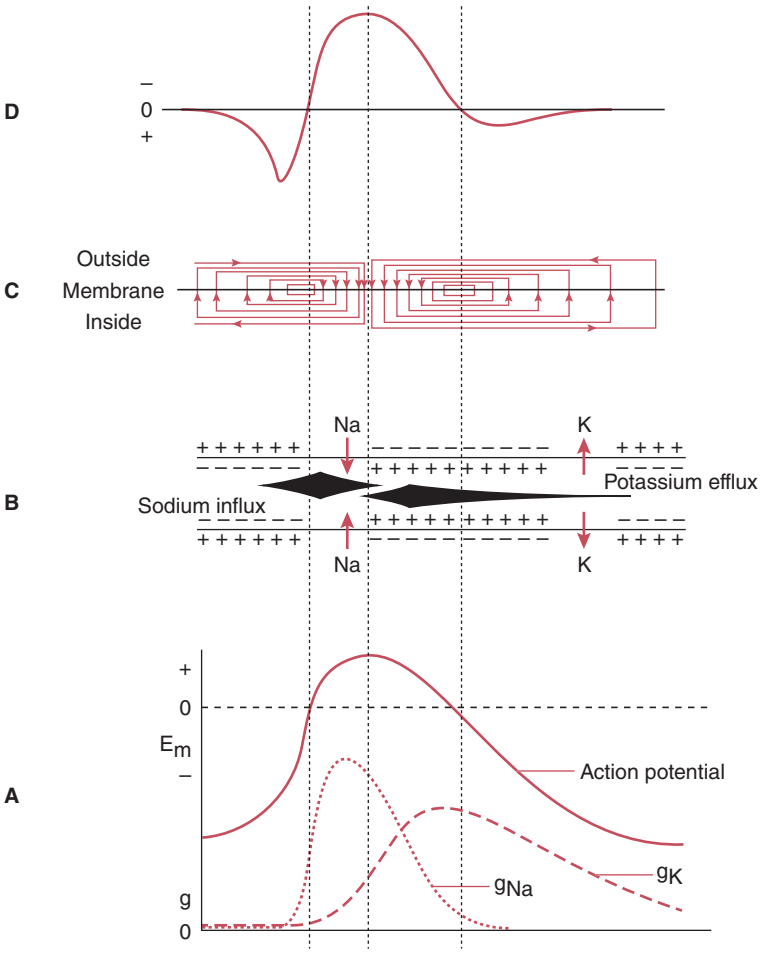


FIGURE 5–5 (A) Sodium (g_{Na}) and potassium (g_K) ion conductance is depicted over time, resulting in an alteration of the transmembrane potential and creating an action potential. (B) The spatial relationship of the sodium and potassium ion influx during an action potential is schematically depicted. Note the alteration of the transmembrane ionic potential differences corresponding to the depolarization and repolarization. (C) Local circuit currents describe the pathways of extracellular sodium ions entering the cell and then migrating longitudinally within the cell. (D) Triphasic extracellular waveform associated with the intracellular monophasic action potential.

- **Na^+ voltage-gated channels (Figure 5–6):**
  - These are protein channels used for ion exchange. They have activation and inactivation gates that undergo conformational changes from a depolarization.
  - They allow increased Na^+ influx into the cell when activated.

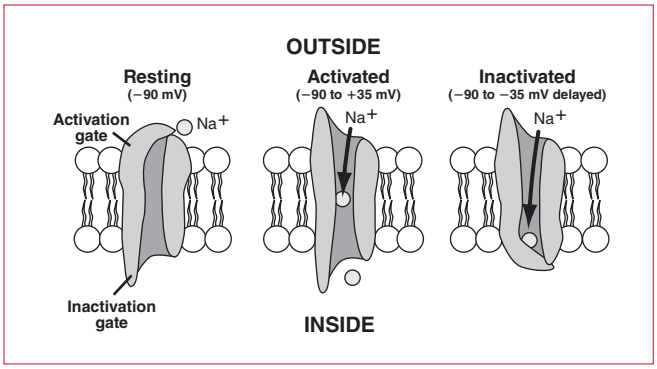


FIGURE 5–6 Na^+ voltage-gated channels.

- **Absolute refractory period:**
  - This pertains to the time after closure of the inactivation gates. They will not immediately reopen. No AP can be formed at this time, no matter how strong a repeated stimulus is used.
  - This may vary in certain disease states (e.g., prolonged in skeletal muscle denervation).
- **Relative refractory period:**
  - This pertains to the period of time after the absolute refractory period. At this time, an AP can be elicited with more intense stimulation. This may also be increased or decreased in certain disease states (e.g., prolonged in skeletal muscle denervation).
- 🕮 **Temperature effects (Figure 5–7):**
  - The Na^+ channels will remain open for approximately 25 µs. A decrease in temperature affects the protein configuration and causes a delay in opening and closing of the gates. This typically changes the waveform appearance, as described later.
  - However, the amplitude can drop due to increased temporal dispersion or phase cancellation.
  - Also, note the difference in focal cooling compared with generalized limb cooling.

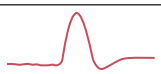
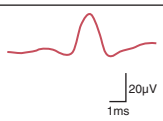
	Temperature	Onset latency (ms)	Peak latency (ms)	Negative spike duration (ms)	Amplitude (µV)	NCV (ms)
	33°C (normal)	2.1	2.6	1.0	22.0	67
	20°C (focal cooling)	2.4	3.1	1.6	27.5	60
	20°C (generalized cooling)	2.8	3.7	1.5	22.0	50

FIGURE 5–7 Decreased temperature effects.
NCV, nerve conduction velocity.

🕮 WAVEFORM CHANGES DUE TO A DECREASE IN TEMPERATURE BELOW 30°C TO 32°C

PARAMETERS	CHANGE
Latency	Prolonged by 1 ms
Amplitude	Increased by 20%
Duration	Increased
Conduction velocity	Decreased by 10 m/s
Phases	Increased

- **Propagation:**
  - As Na^+ goes into the cell from a depolarization, it moves away from the membrane and spreads the current down a path of least resistance along the length of the axon. The affinity to flow back out through the membrane is low due to the myelin sheath covering. Thus, the potential "jumps" to the next group of Na^+ channels, located between the myelin, to areas called the *nodes of Ranvier*.
  - This process of propagating a current from one node to another is known as *saltatory conduction*.
- **Directional recording:**

Orthodromic Recording
- The action potential is recorded traveling in the direction of its typical physiologic conduction.
- Normal physiologic conduction along *motor* fibers travels away from the spinal cord, whereas nerve impulses from *sensory* fibers travel toward the spinal cord.

Antidromic Recording
- The action potential is recorded traveling in the opposite direction of its typical physiologic conduction.
- An antidromic motor nerve study records action potential impulses traveling toward the spinal cord, whereas an antidromic sensory study records sensory impulses traveling away from the spinal cord.

REPOLARIZATION

- The process of bringing the depolarized membrane back to its resting state. It is dependent on Na^+ channel inactivation and K^+ channel activation.
- **K^+ voltage-gated channels (Figure 5–8):**
  - These are protein channels which, after a slight delay, open from a depolarization. This allows K^+ to move out of the cell to establish a charge equilibrium.
  - A delay exists in channel closure, which results in a membrane with a hyperpolarized state called an *overshoot phenomenon*.
  - This process of *potassium conductance* eventually returns the waveform to its baseline due to the K^+ leak channels restoring the RMP.

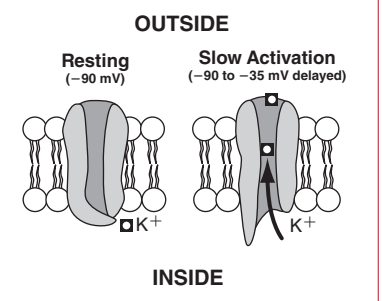


FIGURE 5–8 K^+ voltage-gated channels.

Neuromuscular Junction (Figure 5–9)

- The distal portion of a motor axon has small twig-like terminal branches that innervate individual muscle fibers. This portion of the nerve and single muscle fiber forms the motor endplate.
- The axon terminal, containing various neural structures, including mitochondria and synaptic vesicles with acetylcholine (ACh), does not make direct contact with the muscle fiber. Rather, it remains separate from it by primary and secondary synaptic clefts.

Neuromuscular Junction Components

- **Presynaptic region:**
  - This bulbous area at the axon's terminal zone comprises three storage compartments containing ACh. They are contained in packets called quanta consisting of approximately 5,000 to 10,000 molecules.

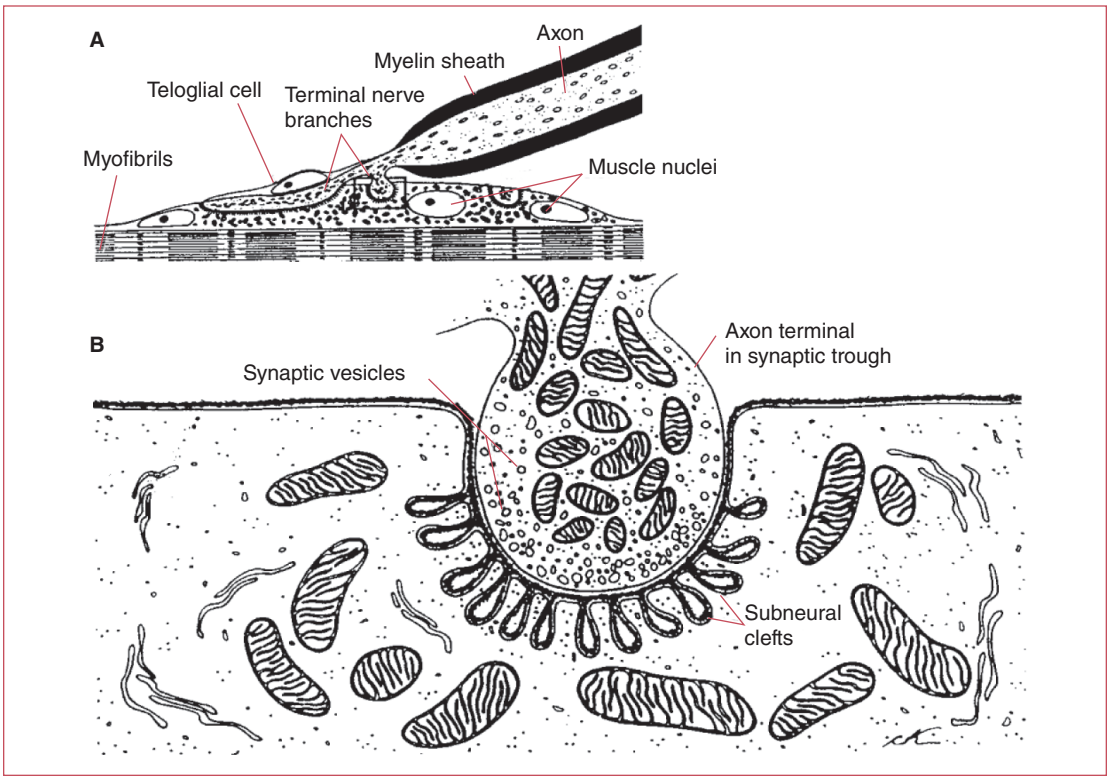


FIGURE 5–9 The neuromuscular junction. (A) Longitudinal section of the junction. (B) Enlarged view.

- The ACh migrates from the main and mobilization storage compartments to replenish the immediate storage compartment, which is depleted in the process of generating each AP. This migration of ACh takes approximately 4 to 5 seconds.

STORAGE COMPARTMENTS	CONTENT
Main store	300,000 quanta
Mobilized store	10,000 quanta
Immediate store	1,000 quanta

- **Synaptic cleft:**
  - This is a space 200 to 500 angstroms wide where ACh crosses from the presynaptic region toward receptors on the postsynaptic region. It contains an enzyme called acetylcholinesterase, which degrades ACh into acetate and choline as it crosses the cleft.
- **Postsynaptic region:**
  - This is a membrane lined with ACh receptors. It has convolutions to increase its surface area by approximately 10× the surface of the presynaptic membrane. At the crests of each fold, receptors are located across from the presynaptic active zones, which are the sites of ACh release. Each postsynaptic ACh receptor requires two molecules of ACh to become activated.

Neuromuscular Junction Physiology

- **Resting state:**
  - During the periods of inactivation, a spontaneous release of a quanta occurs every 5 seconds. This results in production of one miniature endplate potential (MEPP).

- **Excited state:**
  - During the periods of activation, a nerve depolarization opens voltage-gated calcium (Ca^{++}) channels. Ca^{++} floods the nerve terminals and remains there approximately 200 ms.
  - This leads to the release of multiple quanta into the synaptic cleft, which increases the amount of MEPPs. These MEPPs summate to form an endplate potential (EPP), which generates a motor unit action potential (MUAP; Figure 5–10).
- **Safety factor:**
  - The amplitude of an EPP must be high enough to initiate an AP. Normally, the EPP's amplitude is four times the amount needed to initiate an AP. However, the EPP's amplitude drops each time the EPP is created due to a drop in immediate available ACh.
  - This initial excess amplitude of the EPP is called the safety factor and allows time for ACh to move from the main and mobilizing storage compartments to replenish the immediate storage compartment. This avoids a drop of the EPP's amplitude below the threshold needed to cause an AP. The safety factor depends on two parameters:
    - **Quantal content:** Number of ACh quanta released with each nerve depolarization
    - **Quantal response:** Ability of the ACh receptors to respond to the ACh molecules that are released

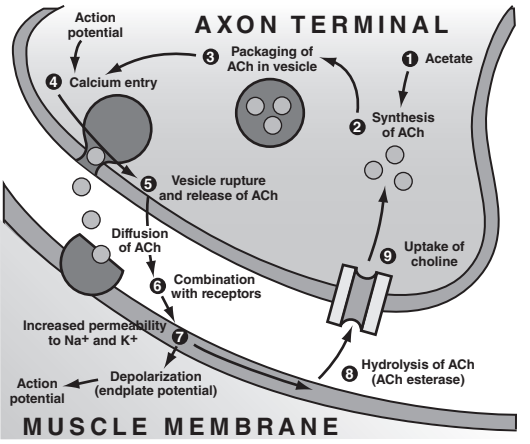


FIGURE 5–10 Acetylcholine release and recycling. ACh, acetylcholine.

Skeletal Muscle Fiber (Figures 5–11 and 5–12)

- This is a cylindrical, multinucleated cell containing contractile elements composed of actin and myosin. The sarcomere is a basic unit of a muscle's myofibril.
- A sarcomere (Figure 5–11) runs from Z-line to Z-line. Its size changes during contraction (Figure 5–12).

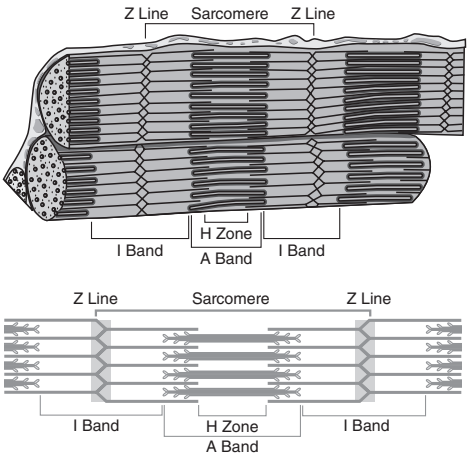


FIGURE 5–11 The sarcomere.

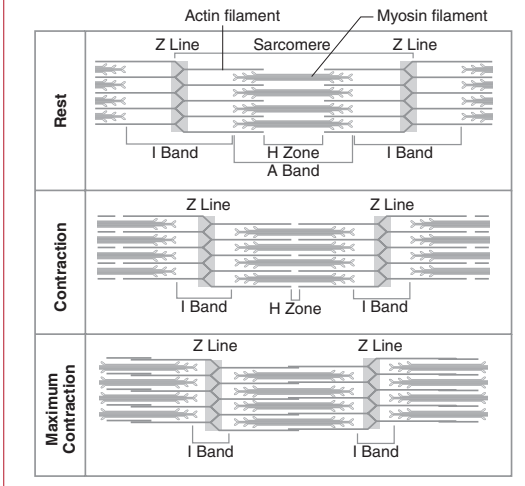


FIGURE 5–12 Sarcomere positional changes.

SKELETAL MUSCLE PHYSIOLOGY

- **Muscle fiber *contraction* (Figure 5–13):**
  - An action initiated by muscle fiber depolarization. The stimulus spreads in both directions on the fiber at 3 to 5 m/s. It penetrates deeper into the muscle through the *T-tubule system*.
  - This causes Ca^{++} to be released from the *sarcoplasmic reticulum*. It binds to the troponin–tropomyosin complex and exposes actin's active sites. Myosin heads, powered by adenosine triphosphate (ATP), bind with the active sites. The actin and myosin filaments slide over each other to shorten the muscle.
- **Muscle fiber *relaxation*:**
  - Powered by ATP, Ca^{++} is actively pumped back into the sarcoplasmic reticulum. This allows the *tropomyosin* to block actin's active sites. Absence of ATP results in rigor mortis due to the actin and myosin filaments remaining permanently joined.

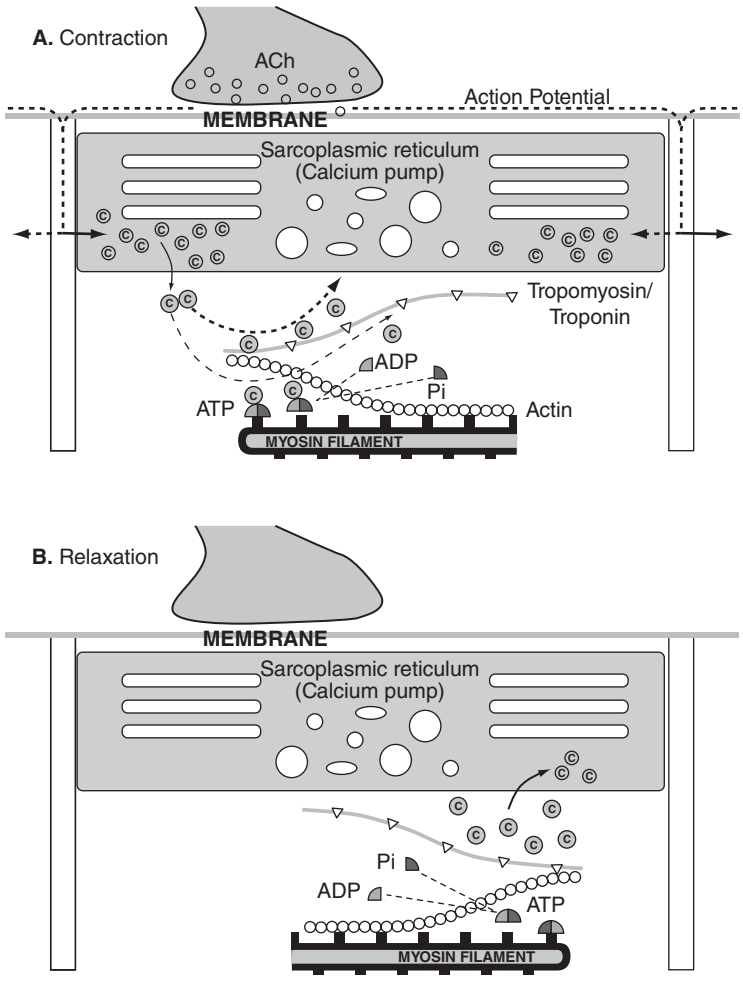


FIGURE 5–13 (A) Muscle contraction and (B) Muscle relaxation: Excitation–contraction coupling in the muscle. This shows an action potential that causes the release of calcium ions from the sarcoplasmic reticulum and then reuptake of the calcium ions by a calcium pump.
ACh, acetylcholine; ADP, adenosine diphosphate; ATP, adenosine triphosphate.

MUSCLE FIBER CLASSIFICATION (TABLE 5–3)

- The characteristics of muscle fibers depend on the motor unit by which it is innervated. If a muscle fiber becomes denervated, it will take on the characteristics of the alpha motor neuron that reinnervates it.

TABLE 5–3 Muscle Fiber Classification

CHARACTERISTICS	TYPE I (SO) SLOW TWITCH OXIDATIVE	TYPE II-A (FOG) FAST TWITCH OXIDATIVE-GLYCOLYTIC	TYPE II-B (FG) FAST TWITCH GLYCOLYTIC
Alpha motor neuron	Small	Large	Large
Color	Dark	Dark	Pale
Recruitment	Early	Late	Late
Fatigue	Highly resistant	Resistant	Sensitive
Effort	Mild (4–8 Hz)	Intermediate (20–30 Hz)	High (20–30 Hz)
Firing frequency	Slow, prolonged	Fast, unsustained	Fast, unsustained
Movements	Fine, precise	Gross	Gross
Innervation ratio	Small	Large	Large
Amplitude/duration	Small	Large	Large
O_2 capacity	Aerobic	Anaerobic	Anaerobic

PATHOPHYSIOLOGY

DEMYELINATION INJURY (FIGURE 5–14)

- This is an injury to the myelin sheath of the nerve, but the axon remains intact. Demyelination increases the membrane capacitance due to the loss of myelin insulation, thus hindering saltatory conduction.
- This translates to slower signal conduction along the axon. The trophic factors of the nerve are maintained, and myelin regeneration is possible due to Schwann cell proliferation. Acutely, conduction block can occur. With time, remyelination can occur. In some chronic disease states, demyelination and remyelination occur repeatedly.

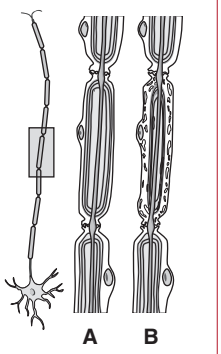


FIGURE 5–14 Demyelination. (A) Normal nerve. (B) Injured segment with myelin breakdown.

Conduction Block
Failure of an AP to propagate past an area of demyelination along axons that are otherwise structurally intact is known as **conduction block**. It can present as a >50% drop in CMAP amplitude between proximal and distal stimulation sites across the area of injury.
AP, action potential; CMAP, compound muscle action potential.

- **Etiologies:**
  - 🕮 Focal compression causing a transient ischemic episode, edema, or myelin invaginations with paranodal intussusceptions (Figure 5–15)
  - Chronic diseases causing degradation of myelin leading to peripheral neuropathies
- **EDX findings of demyelination:**
  - Because demyelination affects the speed of signal conduction along a nerve, it can affect measurements related to time on NCS, notably latency, CV, and temporal dispersion.

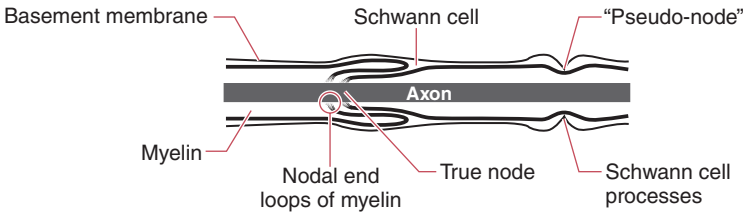


FIGURE 5–15 Paranodal intussusception. Diagram of an invaginating paranode into an adjacent one.

NCS	EMG
Latency: Prolonged Conduction velocity: Decreased Temporal dispersion: Increased Amplitude: May decrease secondary to temporal dispersion and phase cancellation	Normal insertional activity Resting activity: Normal ± myokymia Recruitment: Normal or decreased MUAP: Normal

EMG, electromyography; MUAP, motor unit action potential; NCS, nerve conduction study.

- **Recovery:**
  - Self-limited:
    - The pathology can reverse with cessation of the insulting event. Transient ischemia can be immediately reversed, but edema can take several weeks.
  - Remyelination (Figure 5–16):
    - This is a process of repair in which the demyelinated region develops new myelin produced by the Schwann cells. This new myelin is thinner with shorter internodal distances. CV improves but is usually slower than normal.

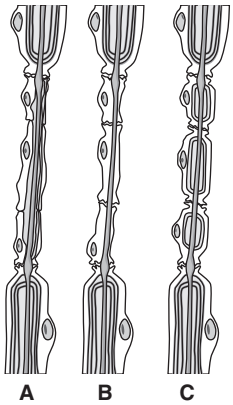


FIGURE 5–16 Remyelination. (A) Myelin digestion and Schwann cell proliferation. (B) Myelin is removed. (C) Remyelination is complete.

AXONAL INJURY (FIGURES 5–17 AND 5–18)

- An injury to the axon may present in one of two typical forms: Axonal degeneration or Wallerian degeneration. Both of these can affect the cell body and cause a central chromatolysis.
- **Axonal degeneration (Figure 5–18):**
  - A nerve injury that begins in a *"dying back"* fashion and affects the nerve in a length-dependent manner. Degeneration of the axon starts distally and continues proximally.
- **Wallerian degeneration (Figure 5–18):**
  - At the site of a nerve lesion, the axon degenerates distally. The nerve segment proximal to the injury site is essentially intact with some minor dying back at the lesion site 1 to 2 cm.
  - For the distal motor axons, the degeneration is generally complete in 7 days.
  - For the distal sensory axons, the degeneration is generally complete in 11 days.
- **Etiology:**
  - Pathology can occur from: (a) focal crush, (b) stretch, (c) transection, or (d) peripheral neuropathies.

- **EDX findings:**
  - Axonal injuries affect the amplitude on nerve conduction waveform, as it is representative of the fastest axons from a nerve. A decrease in amplitude would represent axonal loss.

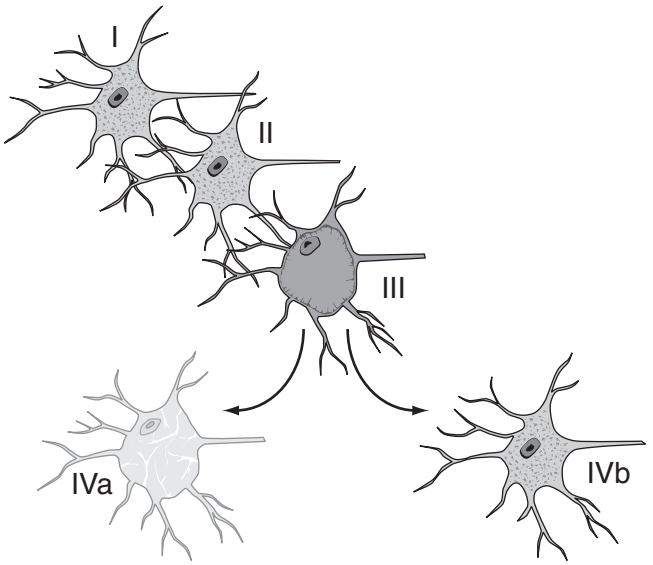


FIGURE 5–17 Axonal injury. (I) Normal nerve cell. (II) Postinjury: Nissl substance degenerates. (III) Swollen cell body with eccentric nucleus. (IVa) Cell death. (IVb) Cell recovery.

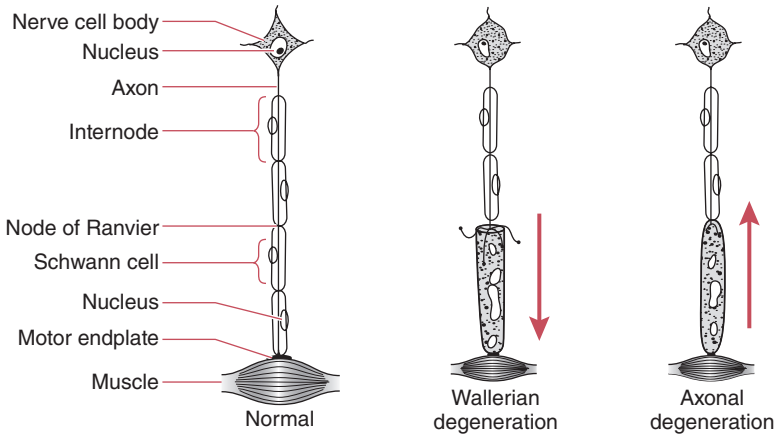


FIGURE 5–18 Schematic representation of axonal injuries.

NCS	EMG
Amplitude: Decreased Temporal dispersion: Normal Conduction velocity and distal latency: Mild slowing of both may occur if the largest and fast conducting axons are lost	Insertional activity: Abnormal Resting activity: Abnormal Recruitment: Decreased MUAP: Abnormal

EMG, electromyography; MUAP, motor unit action potential; NCS, nerve conduction study.

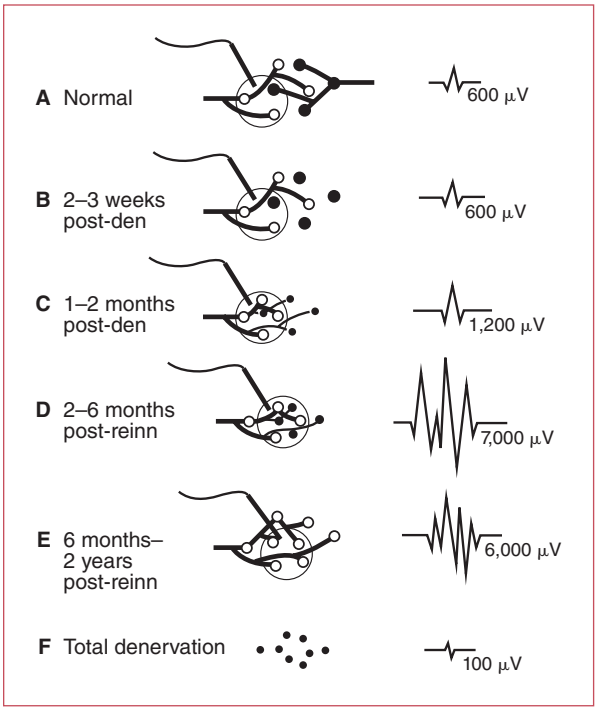


FIGURE 5–19 Motor unit remodeling. (A) Type I: Light circles; Type II: Dark circles. Depolarization of one of the motor units results in a 600 mV MUAP. (B) Following degeneration of the Type II MU at 2 to 3 weeks, the Type I MUAP still yields a 600 mV potential. (C) Within 1 to 2 months, the Type II muscle fibers have atrophied, and collateral sprouting from Type I fibers is beginning to reinnervate them. The Type I motor unit territory has subsequently collapsed due to Type II fiber atrophy, causing a larger MUAP (1,200 mV). (D) As the connections mature, the MUAP demonstrates a further increase in amplitude (7,000 mV) and number of phases. By 6 months, all muscle fibers belonging to the Type I motor units are of the same fiber type; i.e., Type II fibers have been converted to Type I fibers. (E) As maturity continues, the MUAP may decrease its amplitude and phases due to the collaterals conducting potentials more rapidly. (F) An example of complete denervation. MUAP, motor unit action potential.
Source: Copyright ©1995 American Association of Electrodiagnostic Medicine.

- **Recovery:**
  - **Collateral sprouting (Figure 5–19):**
    - This is a process of repair in which a neurite sprouts off the axon of an intact motor unit and innervates denervated muscle fibers of an injured motor unit. The sprouts connect with smaller terminal branches, thinner myelin, and weaker NMJs compared with an uninjured axon. Increased fiber type grouping occurs as muscle fibers become part of the new motor unit and take on its characteristics, increasing the size of its territory. This remodeling results in motor units with poor firing synchronicity, secondary to the immature terminal sprouts. This results in polyphasic waveforms with increased amplitudes.
  - 🕮 **Axonal regrowth (Figure 5–20):**
    - This is a process of repair in which the axon will regrow down its original pathway toward its muscle fibers. It will travel approximately *1 mm/day* or *1 in./month* (*35 mm/month*) if the supporting connective tissue remains intact. These axons will have a decreased diameter, thinner myelin, and shorter internodal distance. With reinnervation, low-amplitude, long-duration, and polyphasic potentials known as *nascent potentials* are formed. If the connective tissue is *not* intact to guide proper nerve regrowth, a neuroma can form with failure to reach the final end organ. Concomitantly, the shorter the distance from injury to end organ, the higher the likelihood for a better prognosis.

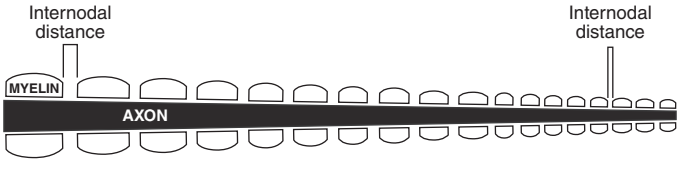


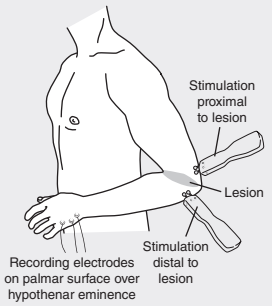
FIGURE 5–20 Axonal regrowth: Axonal diameter is decreased. Myelin is thinner. Internodal distance is shorter.

Collateral Sprouting Versus Axonal Regrowth
If an axon regrows to innervate its original muscle fibers, but collateral sprouting to these fibers has occurred, the nerves possessing the largest axon, thickest myelin, and strongest neuromuscular junction will prevail and keep the muscle fibers.

NERVE INJURY CLASSIFICATION (TABLES 5–4 AND 5–5)

- Two classification systems categorizing nerve injuries are:
  - Seddon classification (Table 5–4)
  - Sunderland classification (Table 5–5 and Figure 5–21)

TABLE 5–4 Seddon Classification

CHARACTERISTICS	NEUROPRAXIA		AXONOTMESIS		NEUROTMESIS	
Etiology	Nerve compression injury		Nerve crush or severe compression injury		Nerve transection injury	
Description	Axon is intact Local myelin injury Conduction block		Axonal interruption Connective tissue/ Schwann cell intact Conduction failure		Axonal interruption Connective tissue disruption Conduction failure	
Nerve conduction studies	The signal is normal with stimulation distal to the lesion but abnormal with stimulation proximal to it.		Conduction resembles neuropraxia for 4–5 days, until Wallerian degeneration occurs.		Conduction initially resembles axonotmesis but does not demonstrate recovery.	
	**Waveform *distal* to lesion:** Immediate 2 weeks >2 weeks	**Waveform *proximal* to lesion:**	**Waveform *distal* to lesion:** Immediate 2 weeks weeks–months	**Waveform *proximal* to lesion:**	**Waveform *distal* to lesion:** Immediate 2 weeks 2 years	**Waveform *proximal* to lesion:**
EMG	Normal/decreased recruitment		Abnormal activity		Abnormal activity	

EMG, electromyography.

TABLE 5–5 Sunderland Classification

TYPE 1	Conduction block (neuropraxia)
TYPE 2	Axonal injury (axonotmesis)
TYPE 3	Type 2 + Endoneurium injury
TYPE 4	Type 3 + Perineurium injury
TYPE 5	Type 4 + Epineurium injury (neurotmesis)

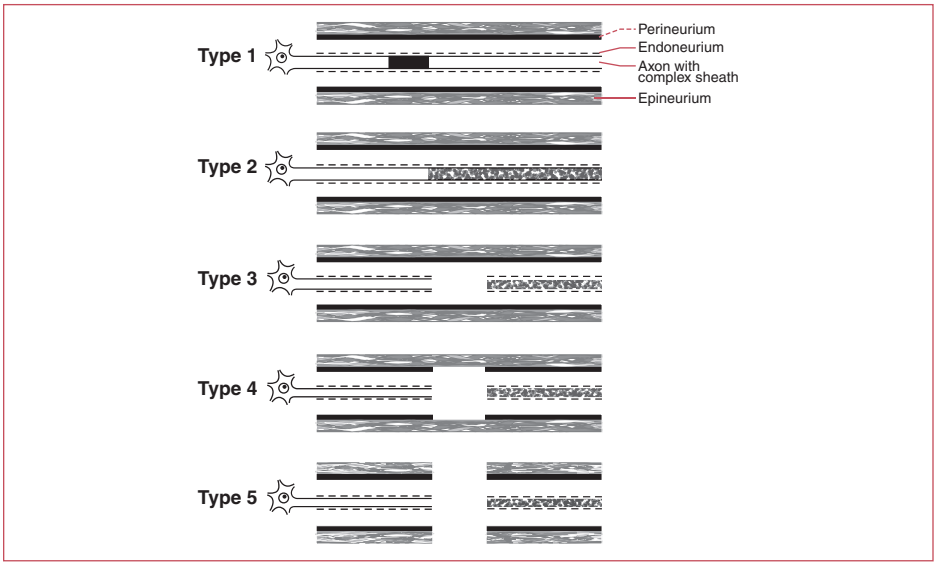


FIGURE 5–21 Sunderland classification.

CLINICAL INSTRUMENTATION

EDX studies consists of NCS and needle EMG.

ELECTRONIC CIRCUITRY (OHM'S LAW)

- An electric current passes through a wire at an intensity of the current **(*I*)** measured in amperes, equal to the voltage **(*V*)** from an electromotor source measured in volts divided by the resistance **(*R*)** measured in ohms. The following formula is known as Ohm's Law:

Current = Voltage/Resistance **(I = V/R or V = I × R)**

ELECTRODIAGNOSTIC INSTRUMENTATION (FIGURE 5–22)

ELECTRODES

These devices are used to record or stimulate the skin surface, muscle, or nerve. An electrode can be an active, reference, stimulating, or ground electrode. They come as either surface or needle electrodes. To obtain a proper reading, the impedance (resistance) between the electrode and skin must be kept low by removing skin lotions, oils, gels, and so on.

- **Recording electrodes:**
  These are devices placed on the skin surface or in the soft tissue to pick up electrical activity from the muscle or nerve. See next section on surface versus needle electrodes for further details.

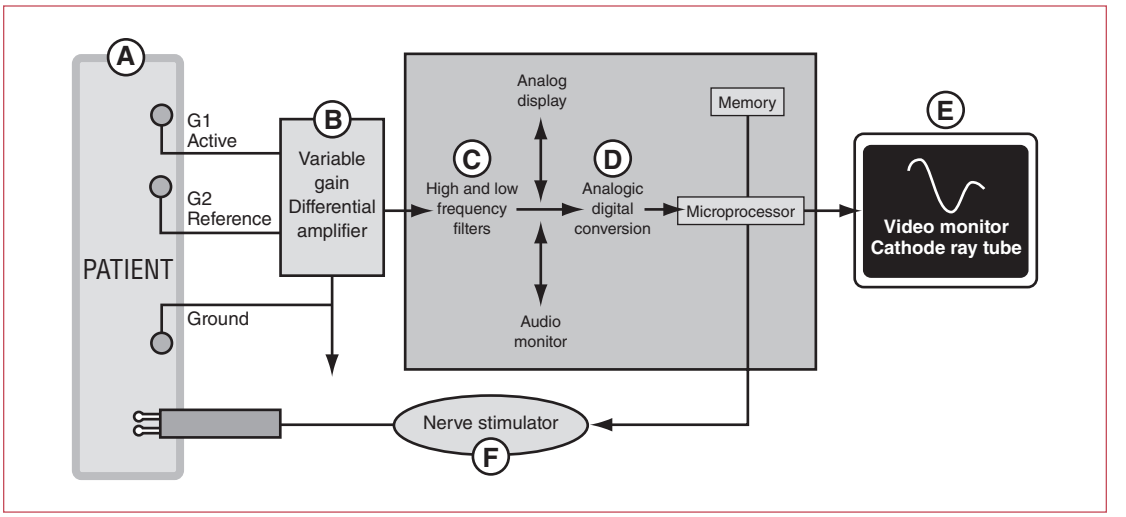


FIGURE 5–22 Electrodiagnostic instrumentation. (A) A patient with recording electrodes has a peripheral nerve excited with the nerve stimulator (F). (B) The differential amplifier receives the action potential. (C) The signal is filtered. (D) The analog signal is converted to a digital representation while being fed to a loudspeaker. (E) The signal is displayed on a cathode ray tube. (F) Stimulator is used to excite the peripheral nervous system.

  - **Active electrode (G1):**
    This pickup records the electrical activity from a nerve actional potential (AP). In a sensory NCS, the recording electrode is placed directly over the nerve, and the electrical activity from the nerve, known as the sensory nerve action potential (SNAP), is recorded. The recording electrode in a motor nerve study is placed over the motor endplate of a muscle that is innervated by that nerve to record the compound muscle action potential (CMAP) signal. The CMAP represents the summation of electrical activity generated by muscle fibers and is an *indirect* representation of electrical activity generated by a motor nerve.
  - **Reference electrode (G2):**
    This pickup is placed over an electrically neutral area (tendon or bone) during a sensory or motor nerve study.
- **Surface electrodes (Figure 5–23):**
  Surface electrodes are placed on the skin to record nerve or muscle APs. They are typically either metal electrodes or disposable electrode stickers lined with an adhesive backing and conductive gel.
- **Needle electrodes:**
  These electrodes are inserted through the skin to record muscle or nerve APs. If used for NCS, the waveform's amplitude and CV cannot be assessed because the needle samples only a few fibers.
  - **Monopolar needle electrode (Figure 5–24):**
    This is a 22- to 30-gauge Teflon-coated needle with an exposed tip of 0.15 to 0.2 mm^2.

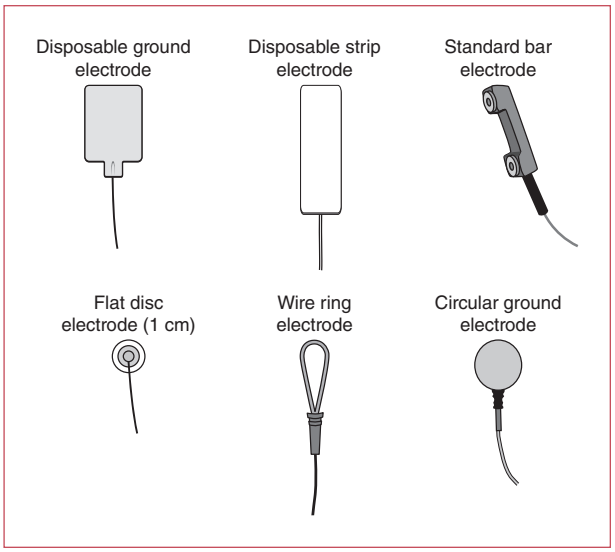


FIGURE 5–23 Various types of surface electrodes.

Advantages:
- Inexpensive
- Conical tip: Omnidirectional recording
- Less painful (Teflon decreases friction)
- Larger recording area (2× concentric)
- Records more positive sharp waves (PSWs) and more abnormal activity in general

Disadvantages:
- Requires a separate needle or surface reference
- Nonstandardized tip area
- Teflon fraying
- May have more interference if the reference is not near the recording electrode

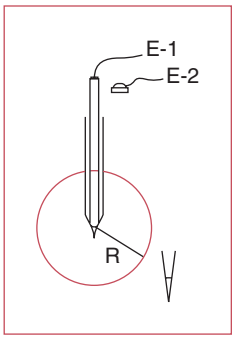


FIGURE 5–24 Monopolar needle electrode.

– **Standard concentric (Coaxial) needle electrode (Figure 5–25):**
This is a 24- to 26-gauge needle (reference) with a bare inner wire (active).

Advantages:
- Standardized exposed area
- Fixed location from reference
- Less interference
- No separate reference
- Used for quantitative EMG

Disadvantages:
- Beveled tip: Unidirectional recording
- Smaller recording area
- MUAPs have smaller amplitudes
- More painful

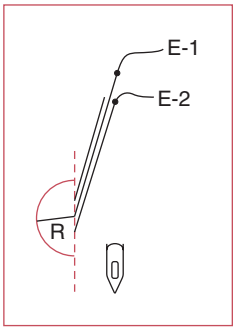


FIGURE 5–25 Concentric needle electrode.

– **Bipolar concentric needle electrode (Figure 5–26):**
This is a needle with the active and reference wires within its lumen.

Advantages:
- Best for isolating MUAP
- Less artifact

Disadvantages:
- Expensive
- More painful

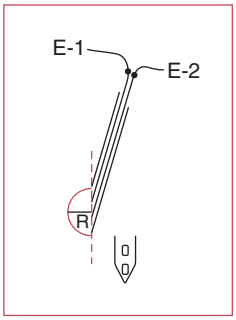


FIGURE 5–26 Bipolar needle electrode.

– **Single-fiber needle electrode (Figure 5–27):**
This is a needle (reference) consisting of an exposed 25-μm diameter wire (active).

Advantages:
- Looks at individual muscle fibers
- Used to assess fiber type density
- Used to assess jitter
- Used to assess fiber blocking
- Helpful in assessing NMJ disorders and motor neuron disorders

Disadvantages:
- Not used for traditional EMG
- Expensive

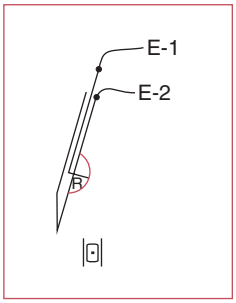


FIGURE 5–27 Single-fiber electrode.

- **Ground electrode:**
This is a zero-voltage, surface reference point placed between the recording electrode and the stimulating electrode.

- **Stimulating electrode (Figure 5–28):**
  This is a bipolar device used to apply an electrical impulse to a nerve to initiate a nerve AP. The stimulator has a cathode and an anode pole:
  - The **cathode** terminal generates a negative impulse that attracts positive charges from the axon.
  - The **anode** terminal generates a positive impulse that attracts negative charges from the axon.

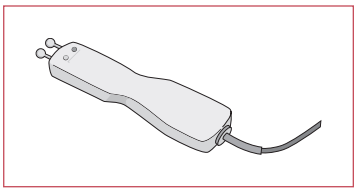


FIGURE 5–28 Bipolar stimulator.

📖 Anodal Block
A theoretical local block that occurs when reversing the stimulator's cathode and anode. This hyperpolarizes the nerve, thus inhibiting the production of an action potential.

NERVE CONDUCTION STUDIES

- NCS are performed by electrically stimulating the nerve and recording the signal. The recorded signal is affected by multiple technical factors, including stimulation intensity and duration, as well as noise and interference signal.

Stimulation Intensity

- **Threshold stimulus:**
  - This is an electrical stimulus occurring at an intensity level just sufficient enough to produce a detectable evoked potential from the nerve.
- **Maximal stimulus:**
  - This is an electrical stimulus at an intensity level in which no further increase in an evoked potential will occur from the nerve with added stimulus intensity.
- **Submaximal stimulus:**
  - This is an electrical stimulus at an intensity below the maximal stimulus level but above the threshold level. This can lead to a falsely lower recorded amplitude and prolonged latency reading, which can give the false impression of an axonopathy or conduction block.
- **Supramaximal stimulus:**
  - This is an electrical stimulus at an intensity at least 20% above the maximal stimulus and is typically used for NCS.
  - 📖 With stimulus intensity set too high, unwanted results may occur due to volume conduction. Volume conduction occurs when the stimulus current spreads through tissue surrounding the nerve. Skin, extracellular fluid, muscles, and other nerves may be stimulated, which can lead to:
    - Decreased conduction times and shortened latencies
    - Altered waveforms
    - Amplitudes remain unchanged

Stimulation Duration

Stimulus duration is usually set at 0.1 ms and may be increased incrementally to ensure supramaximal stimulation. If a monopolar needle is used for stimulation, start at 0.5 ms. Longer stimulus duration will cause more pain.

Stimulation Averaging

This process extracts the desired neurophysiologic signal from larger noise and interference signals. These unwanted signals can occur from biological or environmental sources, such as EMG audio feedback, needle artifact, 60 hertz (Hz) line interference, preamplifier proximity to the machine, fluorescent lights, or the patient.

- 📖 **Signal-to-noise ratio (S:N):**
  The process of *averaging* improves the S:N by a factor that is the square root of the number of averages performed. The number of averages must be increased by a factor of four to double the S:N.

$$\text{S:N} = \frac{\text{Signal amplitude} \times \sqrt{\#\text{ of averages performed}}}{\text{Noise amplitude}}.$$

Stimulus Artifact

This is a defect seen at the time the stimulus is applied to the skin and represents current spread to the electrode. It can be minimized by:

- Placing the ground electrode between the recording electrode and stimulator
- Appropriate anode and cathode placement
- Cleansing the skin from dirt, perspiration, and lotions

DIFFERENTIAL AMPLIFIER (FIGURES 5–22B AND 5–29)

This is a device within a preamplifier that responds to alternating currents of electricity. It cancels waveforms recorded at both the active and reference pickups and amplifies the remaining potentials (Figure 5–29). It should have a high impedance and common mode rejection but low noise from within the system.

Common Mode Rejection Ratio

This refers to selectively amplifying different signals and rejecting common ones. It is usually expressed as dB and should be ≥90 dB. The larger the CMRR, the more efficient the amplifier.

CMRR, common mode rejection ratio; dB, decibels.

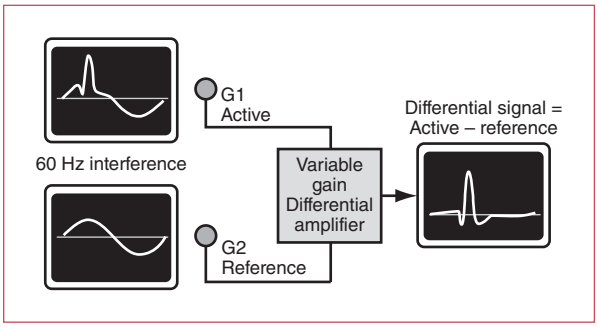


FIGURE 5–29 Schematic representation of differential amplifier function. A differential amplifier only amplifies the difference in the signal present at the active and reference inputs. When 60-Hz interference is the same at both inputs, it is eliminated, leaving only the difference signal, which is the action potential, being measured.

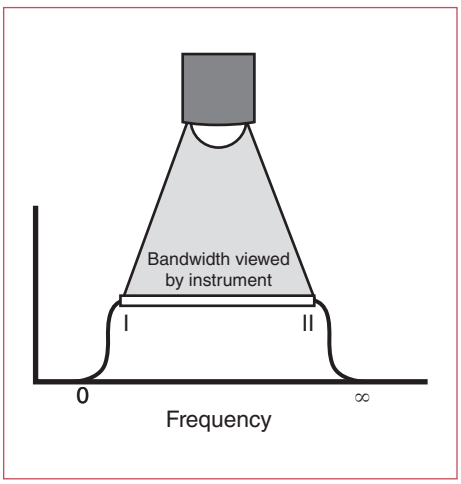


FIGURE 5–30 The frequency bandwidth. This is a schematic representation of the frequencies the filters have allowed the instrument to view. (I) Low-frequency filter. (II) High-frequency filter.

FILTERS (FIGURE 5–30)

This device, composed of resistors and capacitors, functions to exclude unwanted waveforms from being recorded.

- **Types of filters:**
  - **High-frequency (low pass) filter (HFF):** An HFF removes signals with frequencies higher than its cutoff setting. Signals with frequencies lower than (below) the cutoff setting are not affected. This affects the faster portions of the summated waveform.
  - **Low-frequency (high pass) filter (LFF):** An LFF removes signals with frequencies lower than its cutoff setting. Signals higher than (above) the cutoff setting are not affected. This affects the slower portions of the summated waveform.
- **Filter settings:**
  - Sensory NCS: 20 Hz to 10 kHz
  - Motor NCS: 2 Hz to 10 kHz
  - EMG: 20 Hz to 10 kHz

- **Filter adjustments:**
  - Changes in waveforms can be expected with increasing the LFF (e.g., increase from 1 to 500 Hz) or lowering the HFF (e.g., decreasing from 10,000 to 500 Hz, while maintaining the LFF at 1 Hz).

Effects of Filter Changes on Nerve Conduction Study Waveforms

Elevating the Low-Frequency Filter (Figure 5–31 I–IV)
- Reduces the peak latency
- Reduces the amplitude
- Changes potentials from bi- to triphasic
- Does not change the onset latency

Reducing the High-Frequency Filter (Figure 5–32 I–IV)
- Prolongs the peak latency
- Reduces amplitude
- Creates a longer negative spike
- Prolongs the onset latency

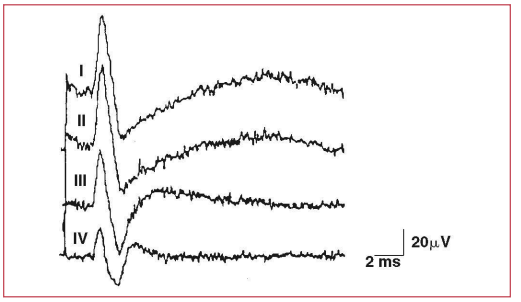


FIGURE 5–31 Elevating the low-frequency filter: Sequential elevation of low-frequency filters (I–IV) from 1 to 500 Hz.

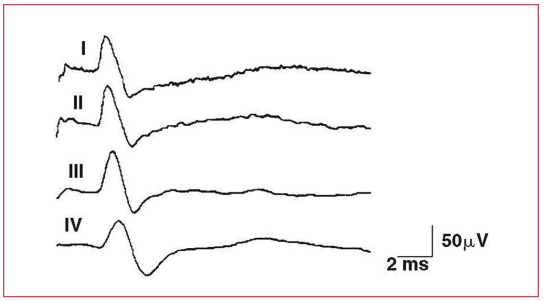


FIGURE 5–32 Reducing the high-frequency filter: Sequential reduction of high-frequency filters (I–IV) from 10,000 to 500 Hz.

SCREEN

- Once a signal has been recorded, amplified, filtered, and passed through, the analog-to-digital converter is displayed on the computer screen. A grid is projected on the screen with the horizontal axis representing *sweep speed* and the vertical axis representing *sensitivity*. Each of these parameters can be adjusted to manipulate the recorded waveform for an accurate measurement.
- **Sweep speed** pertains to the time allocated for each *x*-axis division and is measured in milliseconds.
- **Sensitivity** pertains to the height allocated for each *y*-axis division and is measured in millivolts (mV) or microvolts (μV). The term *gain* is sometimes used interchangeably with sensitivity. Gain is actually a ratio measurement of output to input and does not have a unit value such as mV or μV.
- **Settings:**

	SENSORY	MOTOR	EMG
Sweep Speed	5 ms	2 ms	10 ms
Sensitivity	10 μV	5 mV	100 μV (Insertional activity) 1 mV (Recruitment pattern analysis)

EMG, electromyography.

SAFETY ISSUES

- Each aspect of the EDX exam has certain risk factors. During NCS, electrical risks need to be considered. In needle EMG, certain bleeding risks should be addressed. Though there are no absolute contraindications, these relative risks are weighed against common sense and data obtained in the history (e.g., unexplained bleeding or ecchymosis, cardiac pacemakers, or defibrillators).
- **Electrical risk factors:**
  - Exercise caution in routine EDX studies regarding applying a current to the body. Theoretically, delivering a stimulus may affect factors of cardiac conduction or cause bodily injury from electrical shock.

- **Cardiovascular devices:**
  - Far-field potential generated by routine NCS does not cause electrical activity that would create a detectable stimulation. They pose no risk to implantable pacemakers or intracardiac defibrillators. Yet, a 15-cm (6-in.) separation is suggested between the stimulator and any wires, intravenous (IV) lines, or catheters as a general rule. In addition, one should avoid stimulating the brachial plexus on the same side as a pacemaker or internal cardiac defibrillator.
- **Contraindications:**
  - External cardiac pacemakers: External pacing wires can be electrically sensitive to NCS stimulations.
  - Central line catheters may pose a risk of generating a stimulus in the heart. However, peripheral IV lines are not considered to be problematic.
- **Bleeding risks:**
  - Clinically relevant bleeding issues from an EMG are extremely rare. Considerations to alter the EMG are based on physician comfort for patients taking antiplatelet or anticoagulant medications or with coagulopathies.
  - It is not routinely encouraged to hold anticoagulant or antiplatelet medications for this study. Caution may be exercised in patients with platelet counts <50,000 per microliter and the International Normalized Ratio (INR) has been suggested to be acceptable at 3.0.
- **Other:**
  - Studies at the neck need to take into account the location of the carotid sinus and vagus nerve. Stimulating these could affect the rhythm of the heart.
  - Electrodes should not be placed in a manner where they read a response across the heart.
  - Other common sense issues should be taken into account to avoid external influences on the NCS, such as avoiding contact with exposed metal or wet surfaces.

NERVE CONDUCTION STUDIES

- These studies assess the ability of peripheral nerves to conduct electrical impulses. A representative waveform is generated by nerve stimulation, and its parameters are evaluated to monitor peripheral neuronal function.
  - A sensory nerve actional potential (SNAP) represents the electrical waveform generated by stimulating a peripheral sensory nerve.
  - A compound motor actional potential (CMAP) represents the waveform generated by stimulating a peripheral motor nerve and recording the muscle fiber APs in a muscle.
- **Waveforms:**
  - These recorded potentials represent a compilation of multiple sinusoidal waves. The different phases and amplitudes of each waveform summate or cancel to create a final potential.
  - The final potential displayed represents an average of all the subcomponent's frequencies (Figure 5–33).

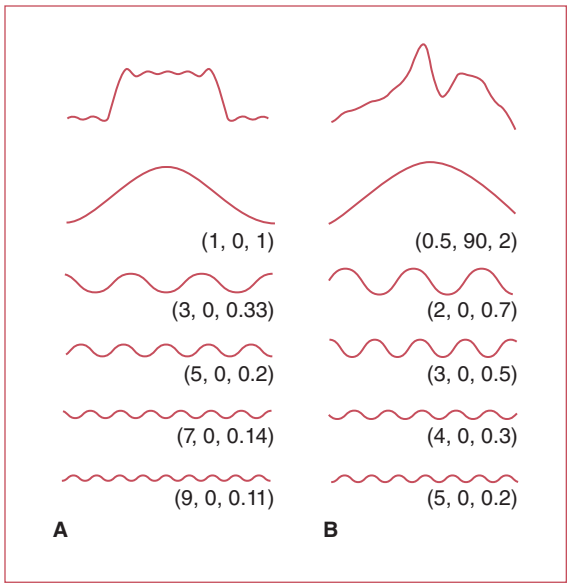


FIGURE 5–33 Waveform subcomponents (summation). (A) A square wave and five subcomponent sine waves, which, when added together, result in the square wave. (B) A more biologic appearing potential and its subcomponent sine waves. The frequency, phase shift, and relative amplitude are described later for each subcomponent waveform.

– Frequency is defined as the number of times the same event occurs in 1 second (cycles per second) and is measured in Hz.

PARAMETERS (FIGURE 5–34)

- **Latency (Table 5–6):**
  - **Onset latency** is the time required for an electrical stimulus to initiate an evoked potential.
    - For sensory studies, onset latencies reflect conduction along the fastest fibers.
    - For motor studies, distal latencies represent the time interval from the motor stimulation site to the NMJ, time delay across the NMJ, and depolarization of the muscle fibers. Conduction along the fastest fibers does not correlate with onset latency.
  - **Peak latency** represents the latency along the majority of the axons and is measured at the peak of the waveform amplitude. Both latencies are primarily dependent on the myelination of a nerve.

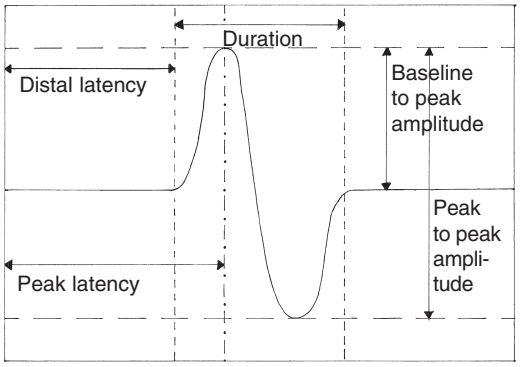


FIGURE 5–34 Waveform parameters.

MEASUREMENTS	DESCRIPTION
Onset latency	• This represents initiation of a conduction response along the fastest axons. • It is recorded at the initial deflection from baseline.
Peak latency	• This represents initiation of conduction along the majority of the axons. – Is a more accurate measure of slowing • It is recorded at the peak of the waveform response.

TABLE 5–6 Nerve Responses of Motor and Sensory Nerve Fibers

NERVE FIBER	NERVE RESPONSE
Motor	**Latency of activation:** This is the time between the initiation of the electrical stimulus and the beginning of saltatory conduction. It is typically 0.1 ms or less. **Conduction:** This is the saltatory conduction of an AP along the myelinated axons to its terminal branches, unmyelinated twigs, and NMJ. **Synaptic transmission:** This is the chemical transmission of the signal across the NMJ to initiate a single-fiber AP. It takes 0.2–1 ms.
Sensory	**Latency of activation:** Same process as earlier **Conduction:** Same process as earlier **Synaptic transmission:** This does not apply due to absence of an NMJ.

AP, action potential; NMJ, neuromuscular junction.

- **CV:**
  - This is the speed an impulse travels along a nerve and is primarily dependent on the integrity of the myelin sheath. It is calculated by dividing the change in distance (proximal stimulation site in millimeter minus distal stimulation site in millimeter) by the change in time (proximal latency in **milliseconds** minus distal latency in milliseconds).
  - *Normal values are* generally *>50 m/s in the upper limbs and >40 m/s in the lower limbs*.
  - It can be decreased with nerve injury and due to technical factors. It should remain normal even in severe axonal injuries, as NCS record the velocity of fastest surviving nerve fibers.

🕮 CONDUCTION VELOCITY VARIATIONS:

Age

- CV for a newborn is 50% that of an adult. At 1 year, it is 80% that of an adult. It is equal to an adult by 3 to 5 years.
- Due to segmental demyelination/remyelination and large fiber loss associated with normal aging, typical changes can be seen.
- After the fifth decade, the CV decreases 1 to 2 m/s per decade.

CV, conduction velocity.

Temperature

- Normal is approximately 32°C for the upper limbs and 30°C for the lower limbs.
- It decreases 2.4 m/s per 1°C dropped.
- A 5% decrease in CV has been described for each 1°C drop below 29°C.

CV, conduction velocity.

- **Amplitude:**
  - This is the maximum voltage difference between two points.
  - In sensory studies, the sensory nerve amplitude reflects the sum of activated sensory nerve fibers and their synchronicity of firing. It is commonly measured from baseline to negative peak, or first negative peak to the next positive peak.
  - In motor studies, the amplitude reflects the number of muscle fibers that have been activated. Recordings are typically measured from baseline to the negative peak. While most cases of reduced CMAP amplitudes are due to a loss of axons (as in a typical axonal neuropathy), other causes of low CMAP amplitude include conduction block, some NMJ disorders, and myopathies.
- **Duration:**
  - This is measured from the initial deflection from baseline to the first baseline crossing.
  - In sensory nerves, it is a measure of synchrony of the sensory nerve fibers firing.
  - In motor nerves, it is a measure of synchrony of the individual muscle fibers firing.
- **Area:**
  - This is a function of both the amplitude and duration of the waveform.
- **Temporal dispersion (Figure 5–35):**
  - This reflects the range of CVs of the fastest and slowest nerve fibers. The waveform spreads out (disperses) with proximal compared with distal stimulation. The area under the waveform remains essentially constant.
  - This is due to slower fiber conduction reaching the recording electrode later than faster fibers.
  - This is not usually seen with more distal stimulation when slow and fast fibers reach the recording electrode at relatively the same time.

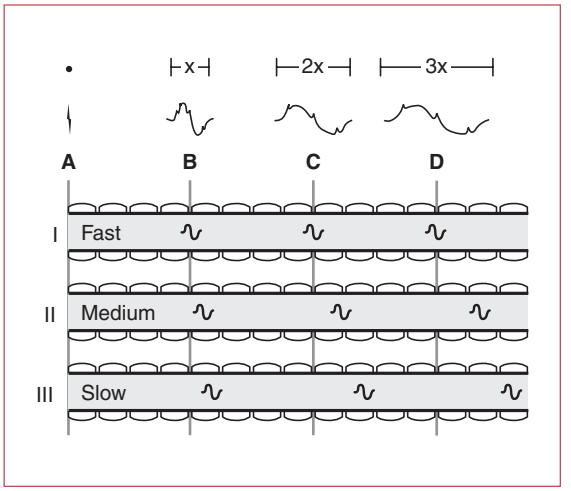


FIGURE 5–35 Temporal dispersion. Three axons of various conduction speed. (I) Fast conduction axon. (II) Medium conduction axon. (III) Slow conducting axon. The signal is measured at different points along the nerve at site A, B, C; then conduction begins at the left and proceeds to the right. At point A, the signal of each axon arrives almost simultaneously, producing a very compact recorded response. At point B, the signals are less well synchronized, producing a smaller amplitude and longer duration response, and this spreading is increased by the time the signals arrive at point C and point D.

- **Phase cancellation: (Figures 5–36 and 5–37):**
  - When comparing a proximal to distal stimulation, a drop in amplitude and increase in duration occur, most notably with a SNAP because of its short duration.
  - When the nerve is stimulated, the APs of one axon may be out of phase with neighboring ones. The negative deflections of one axon can then cancel the positive deflection of another, reducing the amplitude. The summation of these axons creates an AP that appears as one long prolonged wave.
  - For this reason, a drop of 50% is considered normal when recording a proximal SNAP.
  - The CMAP does not have as much of a drop in amplitude because it has a longer duration waveform, and also because of NMJ cushioning. Thus, a smaller decrease in amplitude of approximately 15% is expected.

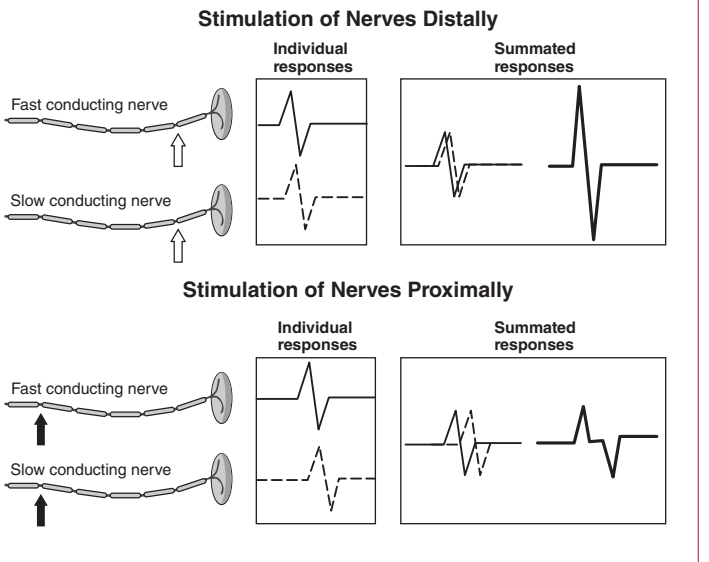


FIGURE 5–36 Sensory—SNAP phase cancellation. Open arrows indicate stimulation of the nerve distally; the phases from the individual SNAPs summate. Closed arrows indicate stimulation of the nerve proximally; with the increased distance, the phases separate enough by the time they reach the recording electrodes to summate less or even cancel. SNAP, sensory nerve action potential.

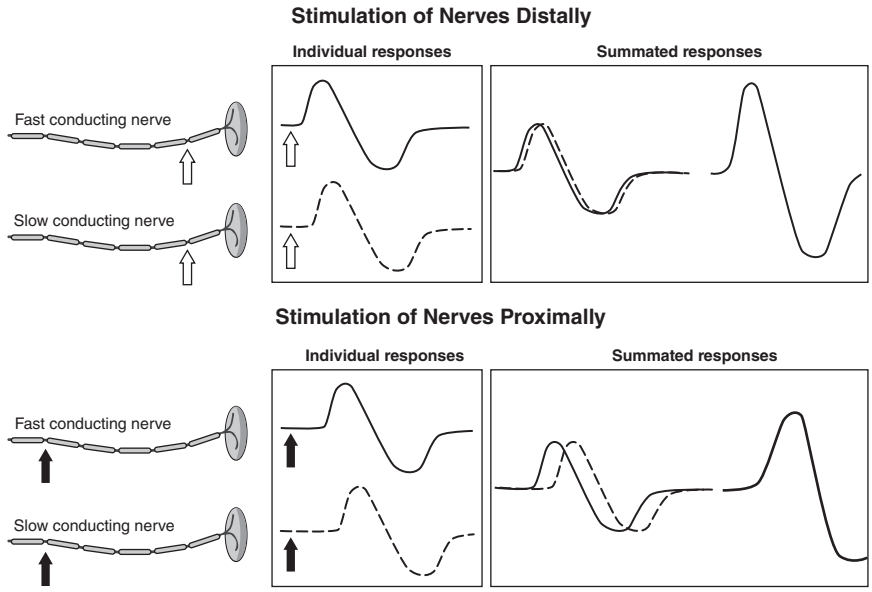


FIGURE 5–37 Motor—CMAP phase cancellation. Open arrows indicate stimulation of the nerve distally, resulting in the discharge of two MUAPs that produce a potential with twice the size. Closed arrows indicate stimulation of the nerve proximally, resulting in two MUAPs that still summate in phase because of the long duration of the MUAPs' negative phases.
CMAP, compound motor action potential; MUAP, motor unit action potential.

SENSORY NERVE ACTION POTENTIAL

- A sensory nerve study represents the conduction of an impulse along the sensory nerve fibers. It can also be useful in localizing a lesion in relation to the DRG (Figure 5–38).
- 🕮 The DRG is located in the intervertebral foramen and contains the sensory cell body. Lesions proximal to it (injuries to the sensory nerve root or to the spinal cord) preserve the SNAP waveform despite clinical sensory abnormalities. This is because axonal transport from the cell body to the peripheral axon continues to remain intact. SNAPs are typically considered more sensitive than CMAPs in the detection of an incomplete peripheral nerve injury.
- **Technical considerations:**
  - **Antidromic studies:**
    - Are easier to record a response than orthodromic studies

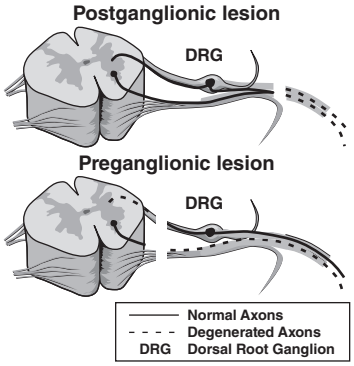


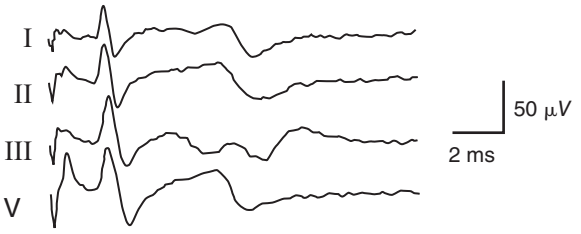
FIGURE 5–38 *Postganglionic injury* results in Wallerian degeneration of both motor and sensory axons. There is physical separation of the axon from the cell bodies in the DRG and the ventral portion of the spinal cord. Compound motor action potential and SNAP responses are diminished or absent. *Preganglionic injury* produces the same injury to the motor fibers but allows the peripheral sensory fibers to remain in contact with their cell body. As a result, SNAPs are normal in this injury.
DRG, dorsal root ganglion; SNAP, sensory nerve action potential.

- May be more comfortable than orthodromic studies due to less stimulation intensity required
- May have larger amplitudes due to the nerve being more superficial at the distal recording sites

– **Recording electrodes:**
- The active and reference pickup should be at least 4 cm apart. Less than this distance will alter the waveform in the following manner (Figure 5–39).

Results When Electrode Separations <4 cm Apart

PARAMETERS	CHANGE
Peak latency	Decreased
Amplitude	Decreased
Duration	Decreased
Rise time	Decreased



Trace	Interelectrode Distance (*cm*)	Amplitude (μ*V*)	Latency (*ms*) Onset	Peak
I	1.0	57	2.8	3.1
II	2.0	73	2.8	3.2
III	3.0	78	2.8	3.4
IV	4.0	87	2.8	3.4

FIGURE 5–39 Active and reference interelectrode distance. Median nerve sensory nerve action potential and the effect of varying the interelectrode separation. (I–IV show a sequential increase in the electrode separation.)

COMPOUND MOTOR ACTION POTENTIAL (FIGURE 5–40)

- Motor nerve studies are known as CMAPs. They are also called M waves. They represent the conduction of an impulse along motor nerve fibers of a motor unit by recording the sum of muscle fiber APs in a muscle.
- It is recorded as an evoked motor potential from a motor point within the muscle. It corresponds to the integrity of the motor unit but cannot distinguish between pre- and postganglionic lesions because the cell body is located in the spinal cord.
- It can be abnormal with normal SNAPs if the lesion is proximal to the DRG (see previous section on SNAPs) or affecting a purely motor nerve (see Figure 5–38).
- **Technical considerations:**
  – **Phases:**
    - The potential should be biphasic with an initial negative deflection. If an initial positive deflection exists, it may be due to:
      - Inappropriate placement of the active electrode from the motor point (Figure 5–41)
      - Volume conduction from other muscles or nerves
      - Anomalous innervations
  – **Recording electrode:**
    - A falsely decreased amplitude and inaccurate latency can be due to:

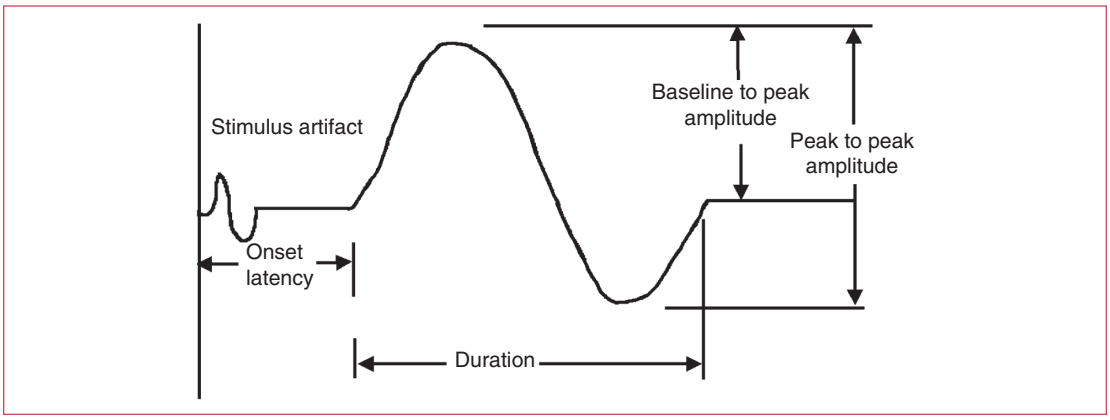


FIGURE 5–40 Compound motor action potential.

- The active and reference electrodes should not be too close together. If this occurs, similar waveforms are recorded at both sites and rejected, decreasing the amplitude of the waveform (Figure 5–42).
- Submaximal stimulation
- Stimulating over thickened skin from callous formation or edema

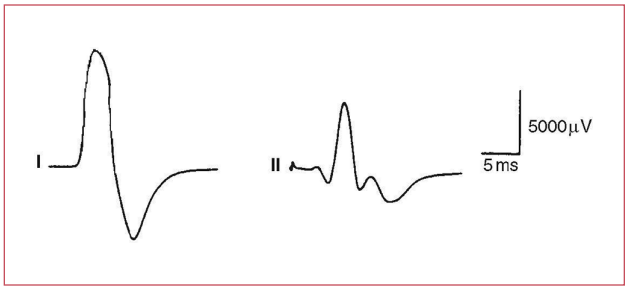
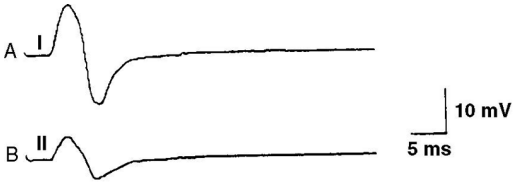


FIGURE 5–41 Compound motor action potential electrode placement. (I) Over the endplate region. (II) Off the endplate region.



Trace	Latency onset	Amplitude: Baseline to peak	Amplitude: Peak to peak
	ms	*mV*	
I	3.4	10	20
II	3.4	4	8

FIGURE 5–42 Compound motor action potential electrode placement. (I) Over the endplate region. (II) Off the endplate region.

MEASUREMENT DIFFERENCES (TABLE 5–7)

TABLE 5–7 Measurement Differences

NCS	SNAP	CMAP
Anatomy examined	Sensory nerve fibers	Motor nerve fibers, NMJ, muscle fibers
Pertinent latencies	Peak or onset latency	Onset latency
Amplitude measurements	Peak to peak (μV)	Baseline to peak (mV)

CMAP, compound motor action potential; NCS, nerve conduction study; NMJ, neuromuscular junction; SNAP, sensory nerve action potential.

CONDUCTION VALUES (TABLE 5–8)

- The following normal values are proposed for the basic motor and sensory NCS. Each EDX lab may have different normal values. Table 5–8 contains proposed normal values for basic motor and sensory NCS.

TABLE 5–8 Normal Values Proposed for Basic Motor and Sensory Nerve Conduction Study

NERVES	VALUES	NERVES	VALUES
Median Motor		**Superficial Radial Sensory**	
Distal latency	3.7 ± 0.3 (8 cm)	Distal latency	2.3 ± 0.4 (10 cm)
Amplitude	13.2 ± 5.0	Amplitude	31.0 ± 20.0
CV	56.7 ± 0.2	CV	58 ± 6.0
Median Sensory		**Peroneal (Fibular) Motor**	
Distal latency	3.2 ± 0.5 (14 cm)	Distal latency	4.5 ± 0.8 (8 cm)
Amplitude	41.2 ± 25.0	Amplitude	4.4 ± 1.4
CV	56.9 ± 4.0	CV below fibula head	51.6 ± 4.1
Ulnar Motor		CV above fibula head	53.9 ± 4.3
Distal latency	3.2 ± 0.5 (8 cm)	**Tibial Motor**	
Amplitude	6.0 ± 1.9	Distal latency	3.4 ± 0.5 (10 cm)
CV below elbow	61.8 ± 5.0	Amplitude	11.8 ± 4.5
CV above elbow	62.7 ± 5.5	CV	53.9 ± 4.3
Ulnar Sensory		**Sural Sensory**	
Distal latency	3.2 ± 2.5 (14 cm)	Distal latency	3.5 ± 0.2 (14 cm)
Amplitude	34.0 ± 12.1	Amplitude	16.6 ± 7.5
CV	57.0 ± 5.0	CV	39.6 ± 2.3
Radial Motor		**Superficial Peroneal Sensory**	
Distal latency	2.4 ± 0.5 (10 cm)	Distal latency	2.9 ± 0.3 (14 cm)
Amplitude	14.0 ± 8.8	Amplitude	20.5 ± 6.1
CV	61.9 ± 5.9	CV	65.7 ± 3.7

CV, conduction velocity.

🕮 H-REFLEX (FIGURE 5–43)

- This NCS late response is an electrically evoked analogue to a *monosynaptic reflex*. In the lower extremity, it is the EDX equivalent of the Achilles reflex.
- It is initiated with a *submaximal stimulus* at a long duration (1.0 ms). This preferentially activates the *IA afferent* nerve fibers, causing an *orthodromic sensory* response to the spinal cord, and then an *orthodromic motor* response back to the recording electrode.
- The waveform can be potentiated with agonist muscle contraction and abolished with antagonist contraction or increased stimulation causing collision blocking.
- 🕮 The morphology and latency remain constant with each stimulation at the appropriate intensity. Note that a mean of 10 F-waves can be used as a surrogate for one H-reflex.

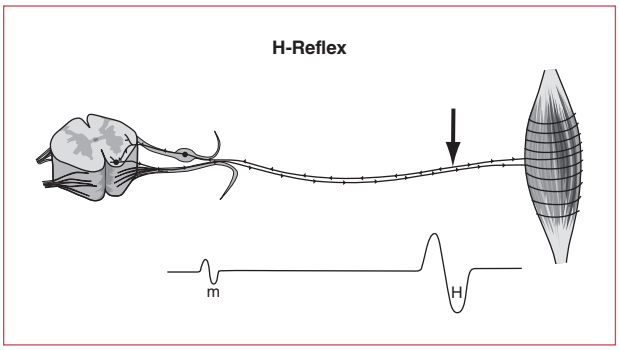


FIGURE 5–43 H-reflex. The H response is obtained by stimulation of the afferent sensory fiber (top) resulting in orthodromic conduction to the spinal cord. In the spinal cord, there is synaptic stimulation of the alpha motor neuron, resulting in the evoked H response in the muscle. A rudimentary M response is produced when a few motor axons are directly stimulated.

- **Function:**
  - It reflects the response of a proximally traveling evoked potential. It is typically used to monitor for an S1 radiculopathy in the lower extremity or a C7 radiculopathy in the upper extremity.
- **Formula:**
  - H-reflex = 9.14 + 0.46 (Leg length in centimeter from the medial malleolus to the popliteal fossa) + 0.1 (age).
- **Latency:**
  - Normal: 28 to 30 ms
  - Side to side difference: >0.5 to 1.0 ms is significant
  - >60 years: Adds 1.8 ms
- **Location:**
  - Soleus muscle: Tibial nerve: S1 pathway
  - Flexor carpi radialis (FCR): Median nerve: C7 pathway
- **Alterations:**
  - This waveform can be seen in all nerves of adults with an upper motor neuron (UMN; corticospinal tract) lesion as well as in normal infants. It is possible to potentiate a waveform by agonist muscle contraction, and inhibit the H-reflex by antagonist contraction.
- **Limitations:**
  - This evaluates a long neural pathway, which can dilute focal lesions and hinder specificity of injury location. It can be normal with incomplete lesions.
  - It also cannot distinguish between acute and chronic lesions. Once it is abnormal, it is always abnormal.
  - Pitfall: While an absent H-reflex can be seen in an S1 radiculopathy, it is NOT a specific finding to diagnose it. Absent H-reflexes can be seen in multiple other conditions, including generalized peripheral neuropathies, plexopathies, and UMN lesions. It can also be a normal finding in elderly adults.

F-WAVE (FIGURE 5–44)

- The F-wave is a small late motor response occurring after the CMAP. It represents a late response from approximately 1% to 5% of the CMAP amplitude. It is produced using a short duration, *supramaximal* stimulation, which initiates an *antidromic motor* response to the anterior horn cells in the spinal cord, which in turn produce an *orthodromic motor* response to the recording electrode.
- The F-wave is a pure motor response and does not represent a true reflex because there is no synapse along the nerve pathway being stimulated. The configuration and latency change with each stimulation due to activation of different groups of anterior horn cells with each stimulation (**Figure 5–45**).

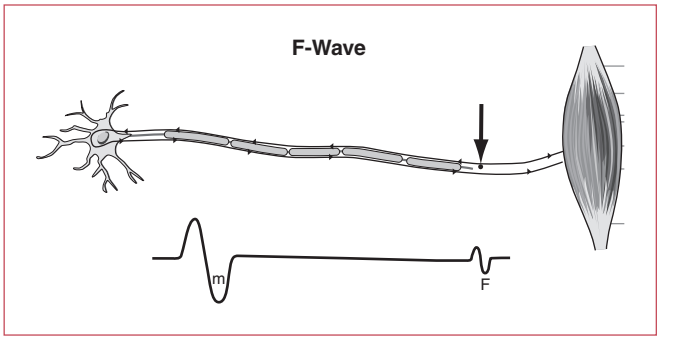


FIGURE 5–44 F-wave response: Stimulation (large arrow pointing to dot) results in depolarization that travels in both directions, first directly to the muscle fiber producing the M response, and retrograde to the axon and to the neuron, where it is repropagated in a small percentage of neurons back down the axons to produce the delayed F-wave response.

- **Function:**
  - May be helpful in polyneuropathies and plexopathies but not overly useful in radiculopathies
- **Latency:**
  - Normal: Upper limb: 28 ms; lower limb: 56 ms
  - Side-to-side difference: 2.0 ms difference in the upper limbs is significant; 4.0 ms difference in the lower limbs is significant.
  - Decreased persistence (occurrence) on repetitive stimulations correlates with a potential abnormality.
- **Location:**
  - It can be obtained from any muscle.
- **Limitations:**
  - This evaluates a long neural pathway, which can dilute focal lesions and hinder specificity of injury location.
  - It only accesses the motor fibers.

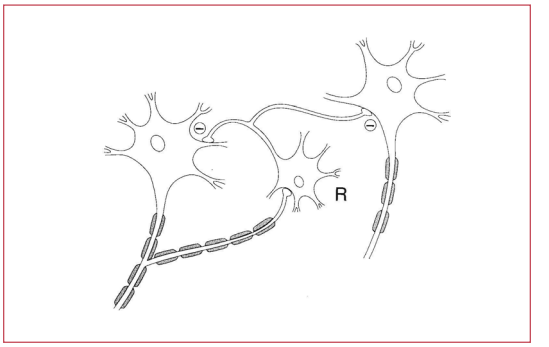


FIGURE 5–45 Renshaw cell activation. Inhibitory neurons, Renshaw cells (R) are activated by a stimulus and in turn suppress (–) firing of the alpha motor neuron.

A-(AXON) WAVE

- When performing a CMAP study, a response can be evoked by a submaximal stimulation and abolished with a supramaximal level. The stimulus can travel antidromically along the motor nerve and becomes diverted along a neural branch formed by collateral sprouting due to a previous denervation and reinnervation process. It typically occurs between the CMAP and F-wave at a constant latency (Figure 5–46).
- **Function:**
  - This waveform represents collateral sprouting following nerve damage.

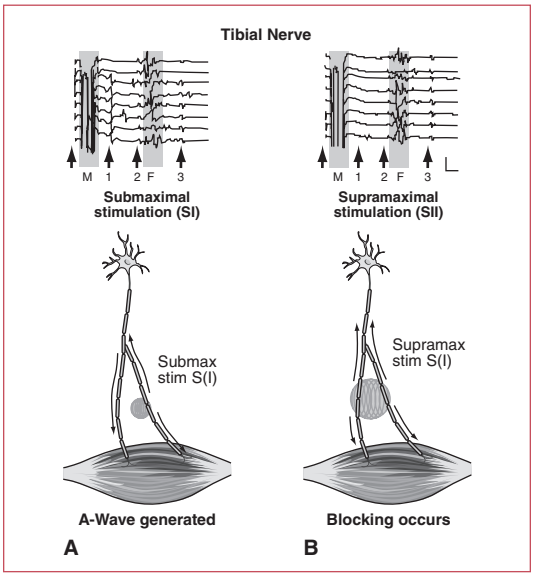


FIGURE 5–46 A-wave. (A) Arrows 1, 2, and 3 represent the A-waves. M is the compound motor action potential and F is the F-wave. S (I) weak stimulus, S (II) strong stimulus. (Note: A-waves seen in S [I] are abolished.) (B) S (I) A-wave generated. S (II) blocking occurs.

BLINK REFLEX (FIGURE 5–47 AND 5–48)

- This NCS is an electrically evoked analogue to the corneal reflex. It is initiated by stimulating the *supraorbital branch* of the trigeminal nerve. The response propagates into the pons and branches to the lateral medulla. It then branches to innervate the ipsilateral and contralateral orbicularis oculi via the facial nerve.
- Two responses are evaluated, an ipsilateral R1 and bilateral R2. The blink is associated with the R2 response (Table 5–9).
- **Latency (Figures 5–49A and 5–49B):**
  - Normal: R1 <13 ms
  - R2 Ipsilateral (direct) <40 ms
  - R2 Contralateral (consensual) <41 ms

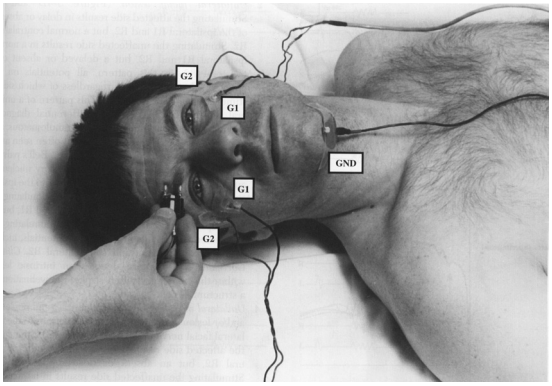


FIGURE 5–47 Blink reflex procedure. Both orbicularis oculi muscles are recorded simultaneously. The active recording electrodes (G1) are placed inferior and slightly lateral to the pupil at mid-position, with the reference recording electrodes (G2) placed just lateral to the lateral canthus. For each side, the ipsilateral supraorbital nerve is recorded over the medial eyebrow. Recording and stimulation sites are shown for a right-sided blink reflex.

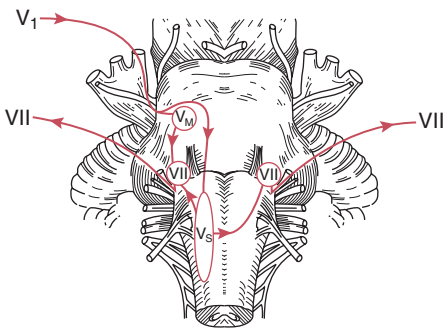


FIGURE 5–48 Blink reflex anatomy. The afferent loop of the blink reflex is mediated by the first division of the trigeminal nerve (V1), which synapses with both the main sensory nucleus of cranial nerve V (VM) in the midpons and the nucleus of the spinal tract of cranial nerve V (VS) in the medulla. The earlier R1 potential is mediated by a disynaptic connection between the main sensory nucleus and the ipsilateral facial motor nucleus (VII). The later R2 responses are mediated by a multisynaptic pathway between the nucleus of the spinal tract of cranial nerve V and both ipsilateral and contralateral facial nuclei (VII). The efferent pathway for both R1 and R2 is mediated via the facial nerve to the orbicularis oculi muscles.

TABLE 5–9 Blink Reflex Pathways

PATHWAY	NERVE FIBERS
Afferent	Sensory branches of CN V (trigeminal nerve)
Efferent	Motor branches of CN VII (facial nerve)

CN, cranial nerve.

RESPONSE	COURSE
R1 (Early)	Through the pons
R2 (Late)	Through the pons and lateral medulla

R1 IS AFFECTED BY LESIONS OF THE:	R2 IS AFFECTED BY:	
Trigeminal nerve	Consciousness level	Valium
Pons	Parkinson's disease	Habituation
Facial nerve	Lateral medullary syndrome	

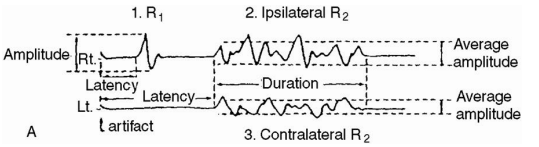


FIGURE 5–49A (A) Blink reflex responses. The pons (1) is the pathway for R1. The pons and lateral medulla (2 and 3) is the pathway for R2.

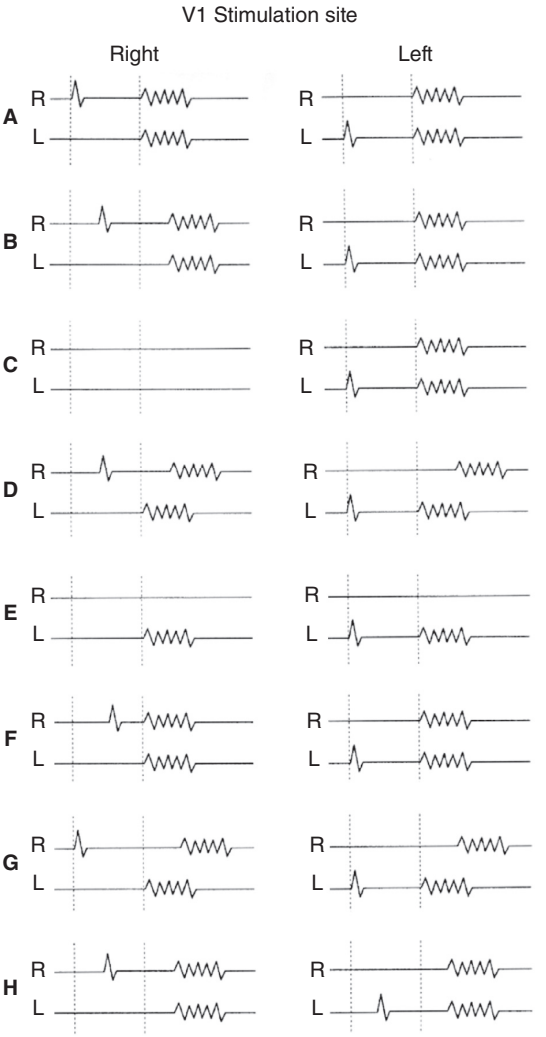


FIGURE 5–49B Blink reflex patterns of abnormalities. (A) **Normal pattern.** Recording both orbicularis oculi muscles; stimulating the supraorbital nerve on each side results in an ipsilateral R1 (early) and bilateral R2 (late) potentials.
(B) **Incomplete right trigeminal lesion.** Stimulating the affected right side, there is a delay of all potentials, including the ipsilateral R1 and R2 and contralateral R2. Stimulating the unaffected side results in all normal potentials.
(C) **Complete right trigeminal lesion.** Stimulating the affected right side, all potentials are absent. Stimulating the unaffected side results in all normal potentials.
(D) **Incomplete right facial lesion.** Stimulating the affected side results in delay of the ipsilateral R1 and R2, but a normal contralateral R2. Stimulating the unaffected side results in a normal ipsilateral R1 and R2, but a delayed contralateral R2. In this pattern, all potentials on the affected side are abnormal, regardless of which side is stimulated.
(E) **Complete right facial lesion.** Stimulating the affected side results in absent ipsilateral R1 and R2 potentials, but a normal contralateral R2. Stimulating the unaffected side results in a normal ipsilateral R1 and R2, but an absent contralateral R2.
(F) **Right midpontine lesion** (main sensory nucleus V and/or lesion of the pontine interneurons to the ipsilateral facial nerve nucleus). Stimulating the affected side results in an absent or delayed R1, but an intact ipsilateral and contralateral R2. Stimulating the unaffected side results in all normal potentials.
(G) **Right medullary lesion** (spinal tract and nucleus V, and/or lesion of the medullary interneurons to the ipsilateral facial nerve nucleus). Stimulating the affected side results in a normal R1 and R2, but an absent or delayed ipsilateral R2. Stimulating the unaffected side results in normal ipsilateral R1 and R2 potentials, but a delayed or absent contralateral R2.
(H) **Demyelinating peripheral polyneuropathy.** All potentials of the blink response may be markedly delayed or absent, reflecting slowing of either or both motor and sensory pathways.

DIRECT FACIAL NERVE STUDY (FIGURE 5–50)

- NCS of cranial nerve VII (facial nerve) is performed by stimulating it distal to the stylomastoid foramen at the angle of the mandible. The response is recorded over the nasalis muscle.
- The patient can present with equal weakness in the upper and lower facial muscles with a peripheral nerve injury.
- If the lesion is rostral to the facial nerve nucleus (central), the lower facial muscles are more severely affected than the upper.

Synkinesis
An aberrant regeneration of axons can occur with facial nerve injuries leading to reinnervation of inappropriate muscles. This may present as lip twitching when closing an eye or crocodile tears when chewing.

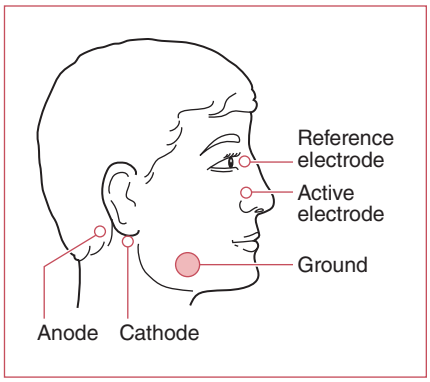


FIGURE 5–50 Facial nerve study.

- **Function:**
  - This monitors for injury to the facial nerve such as seen in Bell's palsy, neoplasms, fractures, middle ear infection, diabetes mellitus, mumps, Lyme disease, and so on.
- **Findings:**
  - This can be monitored periodically over 2 weeks to assess prognosis. Better outcomes are anticipated for demyelinating versus axonal injuries. Absence of an evoked potential in 7 days indicates poor prognosis.
- **Treatment:**
  - Interventions may include prednisone, massage, or electrical stimulation.

Direct Facial Nerve Study

CMAP AMPLITUDE	PROGNOSIS
<10% of the unaffected side	Poor outcome, likely an incomplete recovery. Recovery >1 year.
Between 10% and 30% of the unaffected side	Fair prognosis. Recovery within 2–8 months.
>30% of the unaffected side	Good prognosis. Recovery within 2 months.

CMAP, compound muscle action potential.

PHRENIC MOTOR STUDY

- NCS of phrenic nerve is performed by stimulating just above the clavicle with the cathode and anode pressed posteriorly between the sternal and clavicular heads of the sternocleidomastoid, the recording electrode G1 placed on the xiphoid process and G2 placed at the eighth intercostal space near the costochondral junction.
- The diaphragm CMAP is usually biphasic, with the initial phase upward.
- Needle EMG of the diaphragm:
  - The diaphragm can be studied with a needle EMG electrode, using three approaches: subcostal, lower lateral intercostals, and substernal. The subcostal approach is the most commonly used for the reasons of safety and assurance of proper needle position.
  - In the subcostal approach, insertion of the needle perpendicular to the skin between seventh or eighth rib at the anterior axillary line.
  - It is important to confirm the location of the EMG needle in the diaphragm by listening to rhythmical discharges synchronous with inspiration.
  - The presence of fibrillation potential (FIB) and PSWs is strongly suggestive of neurogenic impairment. In a partially denervated diaphragm, FIBs and PSWs are observed between the bursts of inspiratory electrical activities. In a completely denervated diaphragm, there is absence of inspiratory electrical activity, and only FIBs and PSWs may be seen.
- Treatment for diaphragm paralysis:
  - Phrenic pacing is increasingly being used in patients with central respiratory paralysis and upper cervical spinal cord injury (lesions above C3) to wean them off the ventilators. These patients ideally should not have any intrinsic lung disease.
  - Electrodes can be implanted intrathoracically via thoracotomy and, more recently, with video-assisted thoracoscopic surgery (VATS). Alternatively, electrodes can be placed intramuscularly via a laparoscopic approach. In this approach, intramuscular electrodes are placed near the entrance points of the phrenic nerves using motor-point mapping techniques.

SOMATOSENSORY EVOKED POTENTIALS (FIGURE 5–51)

- This study evaluates time-locked responses of the nervous system to an external stimulus. They represent the function of the ascending sensory pathways using an afferent potential, which travels from the peripheral nerve to the plexus, root, spinal cord (posterior column), contralateral medial lemniscus, thalamus, to the somatosensory cortex.
- It is initiated by a repetitive submaximal stimulation of a sensory nerve, mixed nerve, or dermatome, and is recorded from the spine or scalp. 🕮 The nerves most commonly used for testing are the median for the upper limb and the tibial nerve for the lower limb.
- **Function:**
  - 🕮 Somatosensory evoked potentials (SSEP) monitors for problems such as (a) peripheral nerve injuries, (b) CNS lesions such as multiple sclerosis (MS), or (c) intraoperative monitoring of spinal surgery.
  - Changes in MS are seen 90% of the time, with the lower limb more likely to be abnormal than the upper limb. The most common abnormality is the prolonged interpeak latencies. Amplitude reduction or absence can also be seen.
  - 🕮 During spinal cord surgery, loss of tibial nerve potentials with preservation of median nerve potentials can indicate nerve injury at the level of intervention. Anesthesia will affect SSEP potentials in both the upper and lower limbs.

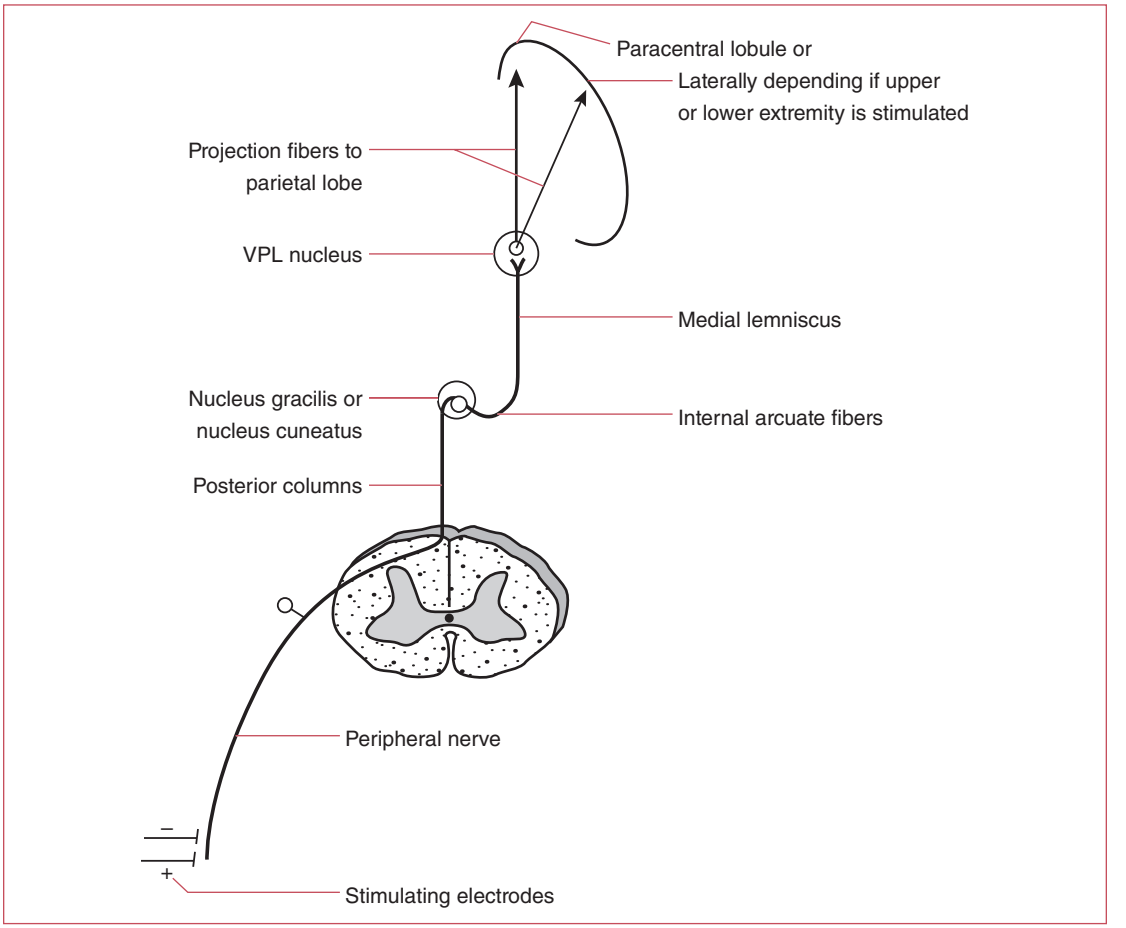


FIGURE 5–51 Somatosensory-evoked potential (SSEP) pathways from the peripheral nerve to the parietal cortex. The fasciculus gracilis is monitored when a lower extremity nerve is stimulated, and the fasciculus cuneatus is monitored when an upper extremity nerve is stimulated.
VPL, ventral posterolateral.

Recording Sites

Median Nerve SSEP (Figure 5–52):

- N9—Erb's point (reflects brachial plexus integrity)
- N11—Roots
- N13—Cervicomedullary junction (nucleus cuneatus)
- P14—Lower brainstem
- N18—Rostral brainstem
- N20—Primary cortical somatosensory receiving area

Tibial nerve SSEP (Figure 5–53):

- PF—Popliteal fossa
- L3—Third lumbar
- N22—T12 and lumbosacral spine
- N45—Cortical

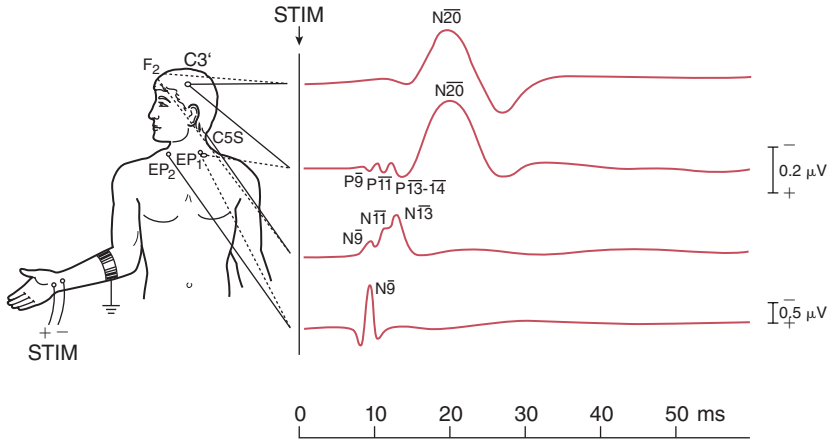


FIGURE 5–52 Median nerve somatosensory evoked potential.

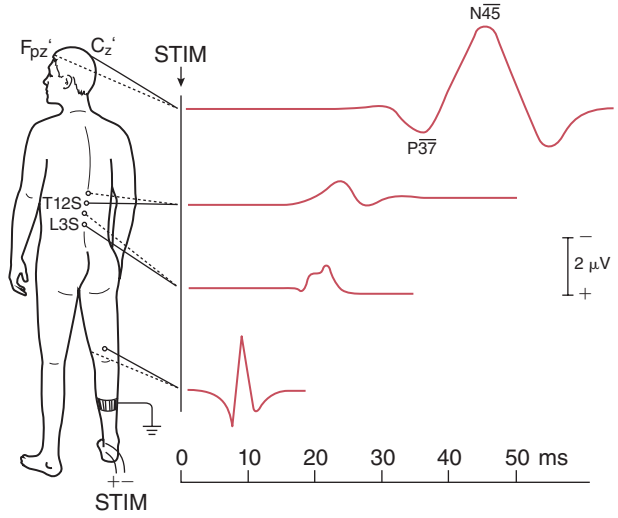


FIGURE 5–53 Tibial nerve somatosensory evoked potential.

SSEP, somatosensory evoked potential.

- **Advantages:**
  - SSEP theoretically evaluates the sensory components of the peripheral nervous system (PNS) and CNS. It can aid in studying disorders of the CNS (brain, brainstem, spinal cord) as well as of dorsal nerve roots and peripheral nerves when severe peripheral disease is noted. Abnormal results present immediately.
- **Limitations:**
  - It only evaluates the nerve fibers sensing vibration and proprioception.
  - It is also limited in its ability to localize a nerve lesion to a focal area. It evaluates a long neural pathway, which may dilute focal lesions and hinder specificity of injury location.
  - It can be adversely affected by sleep and high doses of general anesthetics (halothane, enflurane, isoflurane). This may be avoided with nitrous oxide or low-dose isoflurane.

BASIC NEEDLE ELECTROMYOGRAPHY

- Needle EMG assesses nerve and muscle function. A recording needle electrode is placed into a muscle to evaluate the following parameters:
  - Insertional activity
  - Resting activity
  - Voluntary recruitment

INSERTIONAL ACTIVITY (FIGURE 5–54 AND TABLE 5–10)

- Insertional activity represents discharge potentials that are mechanically provoked by physically disrupting the muscle cell membrane with a needle electrode. This is an electrical injury potential.
- Increased insertional activity may be seen in both neuropathic and myopathic conditions.
- In rare conditions, where significant muscle atrophy has occurred, insertional activity may be decreased.
- Severe, acute ischemia of muscle due to vascular occlusion or compartment syndrome may also produce decreased or absent insertional activity.

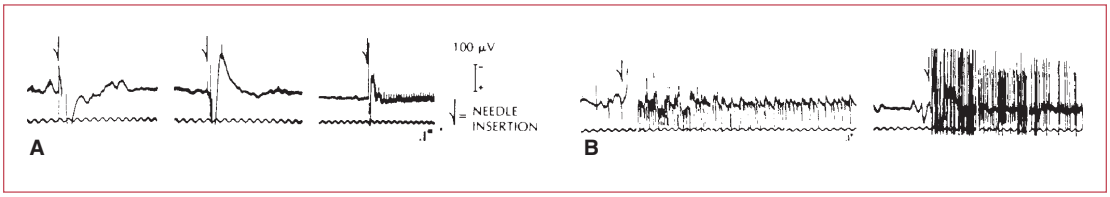


FIGURE 5–54 Insertional activity. (A) Normal. (B) Increased.

TABLE 5–10 Insertional Activity

CHARACTERISTIC	NORMAL	INCREASED	DECREASED
Duration	300 ms	>300–500 ms	<300 ms
Etiology	• Muscle depolarization	• Denervation • Irritable cell membrane	• Fat • Fibrosis • Edema • Electrolyte abnormalities

RESTING ACTIVITY

Normal Spontaneous Activity (Figure 5–55 and Table 5–11)

- After a needle is inserted into a normal muscle, there should be electrical silence at rest. However, if it is placed into or near the NMJ (motor endplate), two waveforms can occur: MEPPs and EPPs. Needle placement in this area is painful, but these waveforms also indicate that the muscle has maintained its innervation.
  - 🕮 MEPPs occur spontaneously at the NMJ, and are referred to as *endplate noise*.
    - They result from the normal spontaneous exocytosis of individual quanta of ACh traveling across the NMJ, leading to a nonpropagated, subthreshold EPP. They have a distinctively small amplitude of 10 to 50 µV and monophasic negative morphology.
    - This "endplate noise" is due to spontaneous quanta release (100–200 quanta), which occurs every 5 seconds. It results in a 10- to 50-µV, nonpropagated potential seen on the screen as an irregular baseline when it is recorded with a standard extracellular needle electrode.
  - EPPs are endplate spikes due to increased ACh release, provoked by needle irritation of the muscle fiber or synchronization of several MEPPs. It results in a propagated single muscle fiber AP.
    - Its hallmark sign is its irregularity, and it always presents as a negative deflection. It can be misinterpreted as a positive wave if the needle is moved a little bit.

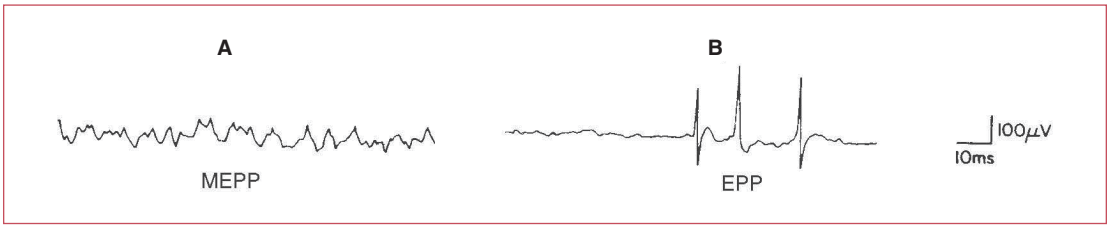


FIGURE 5–55 Endplate activity: (A) MEPP—Monophasic negative potentials primarily present as baseline irregularities because of their small amplitude. (B) EPP—Biphasic, usually negative single-fiber action potential.
EPP, endplate potential; MEPP, miniature endplate potential.

TABLE 5–11 Normal Spontaneous Activity

CHARACTERISTICS	MEPP	EPP
Initial deflection	Negative (monophasic)	Negative (biphasic)
Duration	0.5–1.0 ms	2.0–4.0 ms
Amplitude	10–50 µV	<1,000 µV (1 mV)
Rate	150 Hz	50–100 Hz
Rhythm	Irregular	Irregular
Origin	Endplate	Endplate/provoked
Sound	Sea shell murmur	Sputtering fat in a frying pan

EPP, endplate potential; MEPP, miniature endplate potential.

Abnormal Spontaneous Activity

- Pathologic waveforms can be generated from a muscle fiber or from the motor unit (neural source). They can be caused by a variety of conditions, including muscle denervation or myopathies (inflammation, dystrophies). If originating from a muscle source, the activity can represent lack of muscle fiber innervation.

- The nerve RMP becomes less negative and unstable, causing it to approach the threshold more easily to activate an AP. It either fires independent of external stimulation or is induced by needle movement. If neural in origin, it can have the appearance of MUAPs (Figure 5–56).
- Abnormal spontaneous activity from **muscle fibers** includes:
  - FIBs
  - PSWs
  - Complex repetitive discharges (CRDs)
  - Myotonic discharges
- Abnormal spontaneous activity from a **motor unit** includes:
  - Fasciculation potentials
  - Myokymic discharges
  - Neuromyotonic discharges

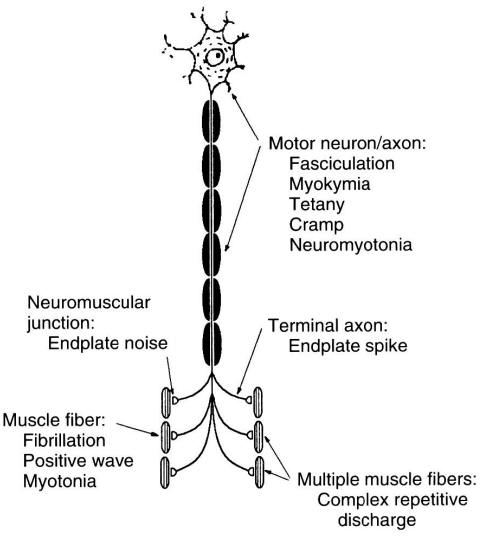


FIGURE 5–56 Spontaneous waveform source generators. Spontaneous activity originates from a variety of source generators. Each generator is associated with a specific morphology.

1. ABNORMAL SPONTANEOUS ACTIVITY GENERATED FROM MUSCLE FIBER OR NERVE PATHOLOGY (TABLES 5–12 AND 5–13)

- **Fibrillations (FIBs) (Figure 5–57):**
  - Spontaneously firing APs originating from denervated single muscle fibers secondary to uncontrolled ACh release.
  - Although they are typically associated with neuropathies, they can also be seen in myopathies.
  - On needle EMG, hallmarks are sounding like "rain on a tin roof" and their regularity of firing.
- **PSWs (Figure 5–58):**
  - Similar to FIBs, PSWs represent spontaneous firing of single muscle fibers.
  - It is a needle recording of an AP of a single muscle fiber. There is propagation to but not past the needle tip. This inhibits the display of the negative deflection of the waveform.
  - Same pathologic significance as FIBs.
  - Hallmarks on EMG are sounding like dull thuds or pops and is also their regularity of firing.
  - Similar to FBIs, they can be found in neuropathies as well as myopathies.

TABLE 5–12 Characteristics of Fibrillations and Positive Sharp Waves

CHARACTERISTICS	FIBs	PSWs
Initial deflection and morphology	Positive (biphasic)	Positive (biphasic)
Duration	1–5 ms	10–30 ms
Amplitude	Early: >300 μV; Late: <25 μV	<1 mV
Rate	1–10 Hz	1–20 Hz
Rhythm	Regular	Regular
Origin	Postjunctional	Postjunctional
Sound	Rain on a tin roof	Dull thud or chug

FIBs, fibrillation potentials; PSWs, positive sharp waves.

TABLE 5–13 Distribution and Intensity of Fibrillation Potentials and Positive Sharp Waves

GRADE	CHARACTERISTICS
0	None
1+	Persistent single runs >1 second in two areas
2+	Moderate runs >1 second in three or more areas
3+	Many discharges in most muscle regions
4+	Continuous discharges in all areas of the muscle

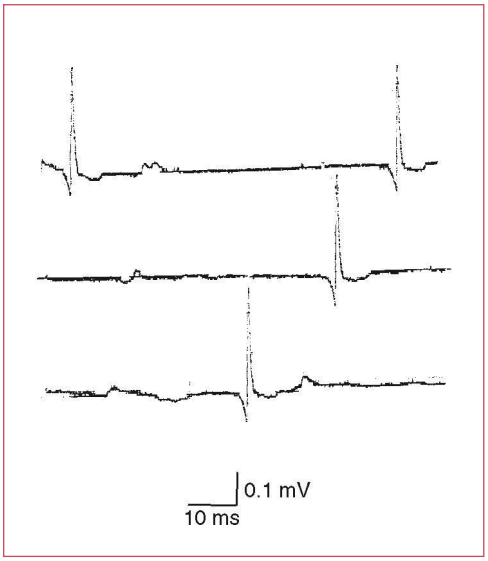


FIGURE 5–57 Fibrillation potentials.

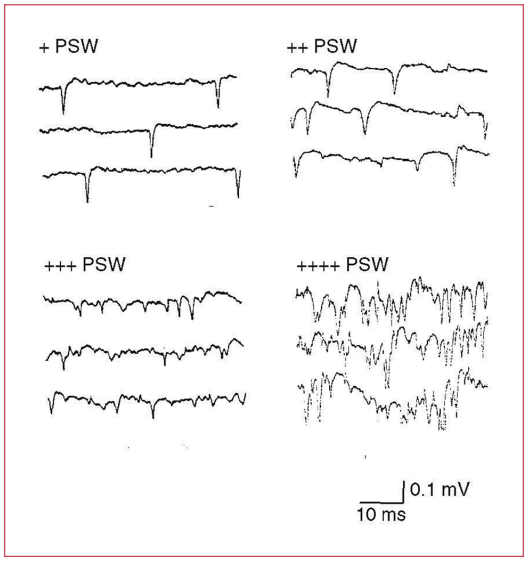


FIGURE 5–58 Positive sharp waves (PSW).

ETIOLOGY:

- *Nerve disorders*: Anterior horn cell disease, radiculopathy, plexopathy, peripheral neuropathy, mononeuropathy
- *NMJ disorders*: Myasthenia gravis, botulism
- *Muscle disorders*: Muscular dystrophies, polymyositis, dermatomyositis, hyperkalemic periodic paralysis, acid maltase deficiency

- **CRDs (Figure 5–59 and Table 5–14):**
  - Formerly known as "bizarre high frequency discharges," CRDs are spontaneous, polyphasic/serrated APs originating from a principal pacemaker, initiating a group of single muscle fibers to fire in near synchrony.

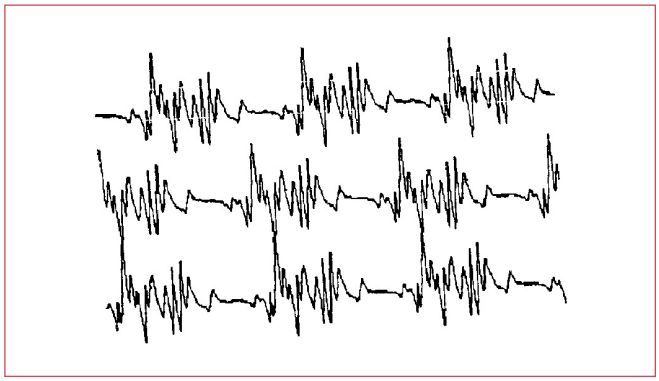


FIGURE 5–59 Complex repetitive discharges.

- The current spreads to the other muscle fibers by *ephaptic transmission* (Figure 5–60). This results from a process in which denervated muscle fibers are reinnervated by collateral sprouting from axons of a neighboring motor unit. When these fibers, in turn, become denervated, a population of muscle fibers now belonging to one motor unit lack neural control. These muscle fibers lie in close proximity to each other and serve as a circuit for the pacemaker fiber.
  - Its hallmark sign is its regular interval between each discharge and within each discharge and a "motor boat engine" sound.

ETIOLOGY

- *Nerve disorders*: Anterior horn cell disease, chronic radiculopathy, and peripheral neuropathy
- *Muscle disorders*: Polymyositis, dermatomyositis, muscular dystrophies, limb-girdle dystrophy, and myxedema
- *Normal variant*

TABLE 5–14 Complex Repetitive Discharges

CHARACTERISTICS	COMPLEX REPETITIVE DISCHARGES
Morphology	Resemble FIBs, PSWs, MUAP
Amplitude	50–1,000 μV
Rate	10–100 Hz
Rhythm	Regular/abrupt start and stop
Origin	Postjunctional/ephaptic transmission
Sound	Motor boat

FIBs, fibrillation potentials; MUAP, motor unit action potentials; PSWs, positive sharp waves.

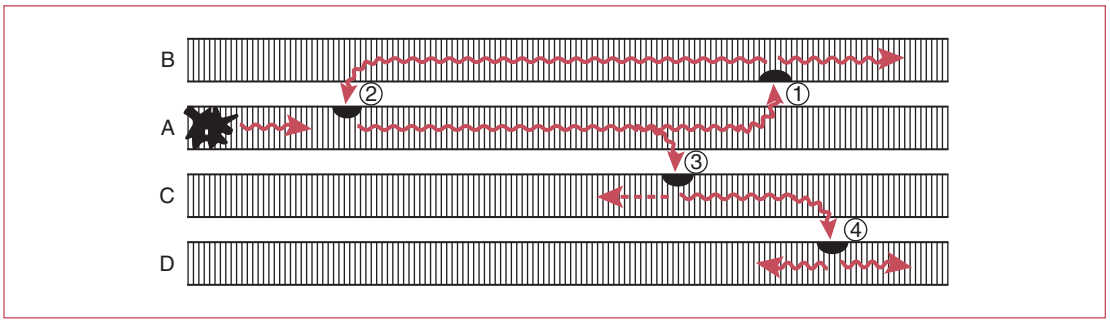


FIGURE 5–60 Ephaptic transmission. Fiber A acts as the pacemaker. It transmits an impulse to B, C, and D, which perpetuates the cycle.

- **Myotonic discharges (Figure 5–61 and Table 5–15).**
  - These are biphasic single muscle fiber APs triggered by needle movement, percussion, or voluntary contraction. They are caused by an alteration of the ion channels in the muscle membrane and can be seen with or without clinical myotonia. Its hallmark sign is the smooth change in rate and amplitude with a "dive bomber" sound.

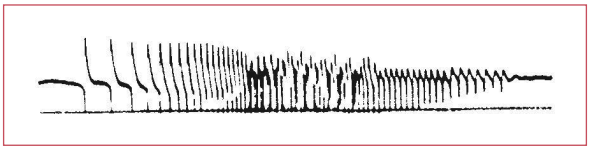


FIGURE 5–61 Myotonic discharges.

TABLE 5–15 Myotonic Discharges

CHARACTERISTICS	MYOTONIC DISCHARGES
Morphology	Resembles: Single-muscle-fiber action potential
Duration	>5–20 ms
Amplitude	20–300 μV
Rate	20–100 Hz
Rhythm	Wax and wane
Origin	Postjunctional
Sound	Dive bomber

ETIOLOGY

- *Nerve disorders*: Chronic radiculopathy, peripheral neuropathy
- *Muscle disorders*: Myotonic dystrophy, myotonia congenita, paramyotonia, polymyositis, dermatomyositis, acid maltase deficiency, and hyperkalemic periodic paralysis
- *Medications*: Propranolol

2. ABNORMAL SPONTANEOUS ACTIVITY GENERATED FROM THE MOTOR UNIT (NEURAL SOURCE)

- **Fasciculations (Figure 5–62 and Tables 5–16 and 5–17):**
  - These are spontaneous discharges originating from the motor neuron or its axon prior to its terminal branches, and result in intermittent muscle fiber contraction. If associated with FIBs or PSWs, they are considered pathologic.
  - A fasciculation has the appearance of a MUAP with an irregular firing pattern. Its hallmark sign is a much slower frequency with a "corn popping" sound. Fasciculations can be pathologic or benign. Differentiating between the two on testing alone is nearly impossible. However, benign fasciculations are not associated with muscle weakness, atrophy, or reflex abnormalities.

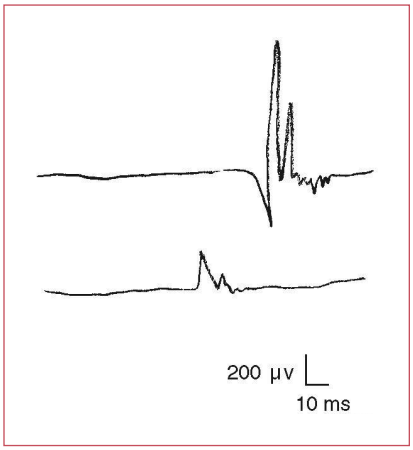


FIGURE 5–62 Fasciculations.

TABLE 5–16 Fasciculations

CHARACTERISTICS	FASCICULATIONS
Morphology	Resembles MUAPs with long, irregular intervals
Duration	5–15 ms
Amplitude	<300 μV
Rate	<1–2 Hz
Rhythm	Irregular
Origin	Prejunctional
Sound	Corn popping—dull, irregular pops

MUAP, motor unit action potential.

TABLE 5–17 Distribution and Intensity of Fasciculations

GRADE	CHARACTERISTIC
0	None
+/–	Equivocal
1+	In two areas, 2–10 per minute
2+	In many areas, 10–15 per minute
3+	All areas, <60 per minute
4+	All areas, >60 per minute

ETIOLOGY

- *Nerve disorders*: Anterior horn cell disease, tetany, Creutzfeldt–Jakob syndrome, radiculopathy, and mononeuropathy
- *Metabolic disorders*: Thyrotoxicosis, tetany
- *Normal variant*

- **Myokymic discharges (Figure 5–63 and Table 5–18):**
  - These are rhythmic groups of MUAPs firing repetitively. They can be associated with clinical myokymia, which presents as slow continuous muscle fiber contractions. This gives a rippling appearance to the overlying skin.
  - Myokymic discharges are seen in multiple conditions but are most commonly seen in radiation-induced neuropathy.
  - Its hallmark sign is the semiregularity between each discharge and within each discharge with the characteristic sound of "marching soldiers."

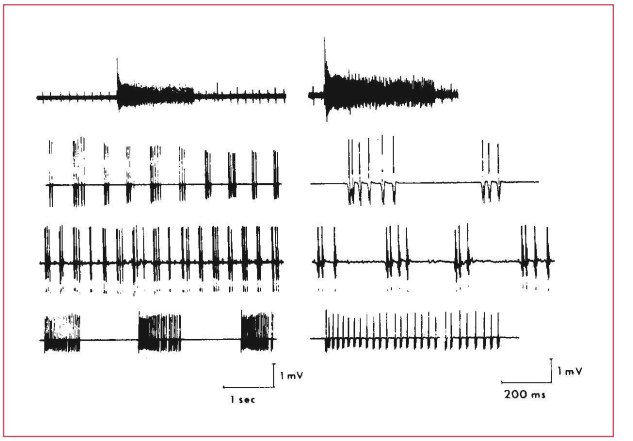


FIGURE 5–63 Myokymic discharges.

ETIOLOGY

- *Facial myokymia*: MS, brainstem neoplasm, polyradiculopathy, and Bell's palsy
- *Extremity myokymia*: Radiation plexopathy (most common), compression neuropathy, and rattlesnake venom

TABLE 5–18 Myokymic Discharges

CHARACTERISTICS	MYOKYMIC DISCHARGES
Morphology	Bursts of MUAPs
Amplitude	100 μV–2 mV
Rate	Discharge: 40–60 Hz; Interdischarge: 0.1–10 Hz
Rhythm	Semiregular
Origin	Nerve
Sound	Marching soldiers

MUAP, motor unit action potential.

- **Neuromyotonic discharges (Figure 5–64 and Table 5–19):**
  - These are high-frequency, repetitive discharges with progressively decremental amplitudes originating from damaged peripheral motor axons. The progressive decrement of their waveforms is due to individual muscle fiber fatigue and drop-off.
  - They are classically seen in neuromyotonia (Isaac's syndrome). This is a disorder associated with continuous muscle fiber activity resulting in the appearance of muscle rippling and stiffness secondary to irritable nerves.
  - Hallmark signs on EMG include high-frequency, repetitive discharges with progressively decremental amplitudes with a characteristic "pinging" sound on EMG.

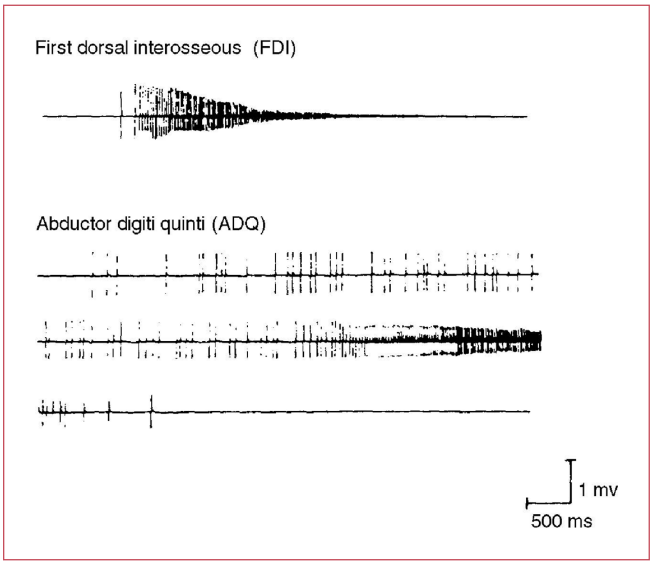


FIGURE 5–64 Neuromyotonic discharges in spinal muscular atrophy.

ETIOLOGY

- *Nerve disorder*: Neuromyotonia, chronic neuropathic disease, and tetany
- *Toxins*: Anticholinesterase

TABLE 5–19 Neuromyotonic Discharges

CHARACTERISTICS	NEUROMYOTONIC DISCHARGES
Morphology	High-frequency, repetitive discharges with progressively decremental amplitudes
Amplitude	Progressively decremental
Rate	150–250 Hz
Rhythm	Regular; discharges continuously or in bursts
Sound	"Pinging"
Appearance	Tornado-like

3. CRAMP DISCHARGES (FIGURE 5–65 AND TABLE 5–20)

- These are spontaneous discharges associated with repetitive firing of MUAPs in a large area of muscle. They originate from high-frequency discharges from motor axons and are not muscle in primary origin.
- They are usually associated with painful, involuntary muscle contractions and are synchronous.

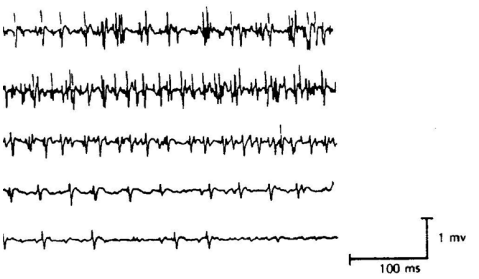


FIGURE 5–65 Muscle cramp discharges.

TABLE 5–20 Cramp Discharges

CHARACTERISTICS	CRAMP DISCHARGES
Morphology	Multiple, normal MUAPs firing at a high, irregular frequency
Duration	Abrupt onset and cessation
Amplitude	Up to 1 mV
Rate	40–60 Hz
Rhythm	Irregular

MUAP, motor unit action potential.

ETIOLOGY

- Electrolyte disturbances, uremia, pregnancy, myxedema, strenuous exercises, prolonged muscle contraction, liver cirrhosis, myotonia congenita, myotonic dystrophy, and stiff-man's syndrome.

4. ARTIFACT POTENTIALS (FIGURES 5–66 AND 5–67)

- These are waveforms that obscure the neurophysiologic signals. Interference potentials are unwanted signals occurring from outside the system being studied.
- Noise is described as unwanted signals occurring from within the system. They arise from the EMG instrument, printer, unshielded power cords, electrical outlets, fluorescent lights, or a pacemaker.

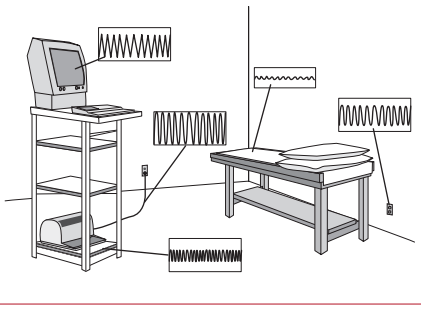


FIGURE 5–66 Source of noise and interference. Noise is greatest near electrical outlets, power cords, and equipment including the electromyography instrument itself.

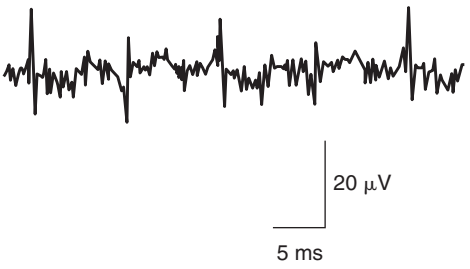


FIGURE 5–67 Fluorescent light interference. *Source*: From Dumitru D. *Electrodiagnostic Medicine*. Hanley & Belfus; 1995, with permission.

EXERTIONAL ACTIVITY

- Voluntary muscle fiber activity that can be electrically recorded and summated as a MUAP.
- **MUAP**
  (Figure 5–68 and Table 5–21):
  - This is a compound AP from muscle fibers belonging to a single motor unit, within the recording range of the needle electrode. Its territory is typically 5 to 15 mm.

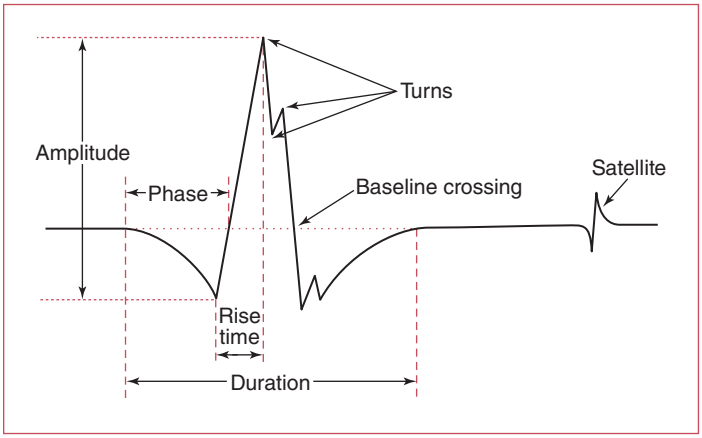


FIGURE 5–68 Motor unit action potential.

TABLE 5–21 Motor Unit Action Potentials

CHARACTERISTICS	MUAP
Initial deflection	Positive/negative
Duration	5–15 ms
Amplitude	1 μV to 2 mV
Rise time	<500 ms
Phases	2–4
Rate	Exertion-dependent
Rhythm	Exertion-dependent
Origin	Prejunctional

MUAP, motor unit action potential.

Parameters

- **Amplitude:**
  - This represents the muscle fibers recorded near the needle electrode. It is measured from the most positive to the most negative peak. It can be increased from a reinnervation process, decreased from loss of muscle fibers, or variable due to blocking associated with NMJ disorders.
  - Normal: 1 mV
- **Rise time:**
  - This represents the time it takes the MUAP to go from its baseline to the peak of the negative wave. It represents proximity of the needle to the motor unit.
  - Normal: <500 μs
- **Duration:**
  - This represents the number of muscle fibers within the motor unit. It is measured from the waveform's initial departure from baseline to its final return. It increases (>15 ms) as the

motor unit territory increases from collateral sprouting or decreases (<5 ms) with the loss of muscle fibers.
  - Normal: 5 to 15 ms
- **Turns:**
  - These are changes in the direction of the waveform that do not cross the baseline. They are also called serrations.
- **Phases:**
  - This represents the synchronicity of muscle fiber APs firing. They are calculated as the number of baseline crossings plus 1.
  - Five or more baseline crossings represent *polyphasicity*. This can result from pathology causing muscle fiber dropout, alterations in fiber CV, or reinnervation from collateral sprouting.
    - However, this can occur normally 15% (concentric needle) or 30% (monopolar needle) of the time in adults, or more frequently in the elderly.
  - Normal: 2 to 4

Abnormalities

- **Long-duration, large-amplitude (LDLA) polyphasic potentials:**
  - These potentials typically occur as a result of denervation and reinnervation from collateral sprouting. This reinnervation process causes an increased number of muscle fibers per motor unit.
  - It is most commonly seen in neuropathic diseases but can also occur in chronic myopathic disease from fiber splitting, as seen in an inflammatory or dystrophic myopathy. The recruitment patterns help distinguish between a myopathic or neuropathic process.
- **Short-duration, small-amplitude (SDSA) polyphasic potentials:**
  - These potentials typically occur from a dropout or dysfunction of muscle fibers. They are most commonly seen in myopathic diseases and NMJ disorders. It can also occur from a severe neuropathic injury leading to nascent motor unit potentials resulting from axon regrowth.
  - Again, the two processes are differentiated by their recruitment patterns.
- **Neuropathic potentials:**
  - This term is not considered to be accurate because certain disease processes mimic MUAP features, regardless of whether the pathology originated in nerve or muscle. It refers to LDLA MUAPs that occur from a denervation and reinnervation process typically seen in neuropathies.
  - This terminology also lacks the appropriate quantitative description of the motor unit parameters.
- **Myopathic potentials:**
  - This term is also not considered accurate because certain disease processes mimic MUAP features, regardless of the pathology muscle or nerve in origin. It refers to SDSA MUAPs that occur from muscle disorders.
  - This terminology lacks the appropriate quantitative description of the motor unit parameters.
- **Unstable potentials:**
  - This refers to the occurrence of variations in the MUAP's amplitude, duration, and slope.
  - It is most commonly seen in NMJ disorders, which cause irregular blocking of discharges. It also can occur in motor neuron disorders, neuropathic disorders, or muscle trauma and reinnervation.
- 🕮 **Satellite potentials:**
  - These are small potentials seen in early reinnervation.
  - They are time-locked potentials that trail the main MUAP. It can be due to the newly formed neural sprouting that often is small, unmyelinated or thinly myelinated → very slowly conducting.
  - These satellite potentials are extremely unstable and may vary slightly in their firing rate (FR) or may block and not fire at all. Over time, as the sprout matures and the CV increases, the satellite potential will become an additional phase or serration within the main complex.

- **Doublet/multiple potentials:**
  - This refers to two or more MUAPs firing recurrently and together in a semirhythmic fashion. It is seen in ischemia, hyperventilation, tetany, motor neuron disorder, or metabolic diseases.
- **Giant potentials:**
  - This refers to the extremely large MUAPs (>5 mV) that occur in disease processes such as poliomyelitis. These are also described as large amplitude potentials.

RECRUITMENT (TABLE 5–22)

- Recruitment refers to the ability to add successive motor units to increase the force of a contraction. With normal minimal contraction, a single MUAP fires before a second MUAP is recruited.

TABLE 5–22 **Normal Motor Unit Action Potential Recruitment Pattern**

FIRST (A)	SECOND (B)	THIRD (C)	FOURTH (D)
A—5 Hz			
A—10 Hz	B—5 Hz		
A—15 Hz	B—10 Hz	C—5 Hz	
A—20 Hz	B—15 Hz	C—10 Hz	D—5 Hz

Motor Unit Recruitment Patterns (Figure 5–69)

- **Normal MUAP recruitment pattern—The rule of fives:**
  - The onset frequency of the first MUAP begins at approximately 5 Hz. To generate more force, the FR and recruitment of more motor units must be increased.
  - When the FR reaches approximately 10 Hz, a second MUAP begins at approximately 5 Hz.
  - When the first MUAP reaches a rate of 15 Hz, the second should be at 10 Hz and a third will begin at 5 Hz.
  - As more force is needed, the FR of the first may reach 20 Hz, the second 15 Hz, the third 10 Hz, and a fourth will begin at approximately 5 Hz.

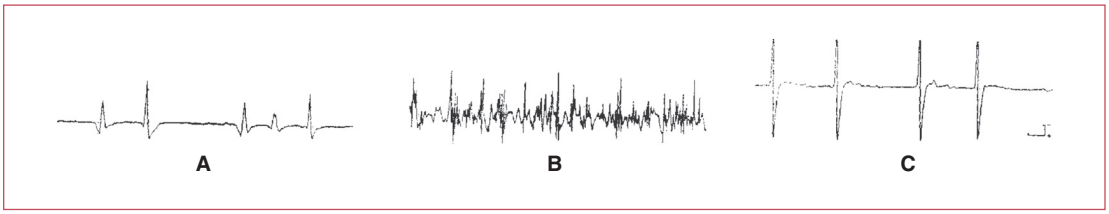


FIGURE 5–69 Motor unit recruitment. (A) Normal. (B) Early (myopathic). (C) Decreased (neuropathic).

- **Early recruitment (myopathic):**
  - Refers to abnormally early firing of many MUAPs with a mild contraction.
  - It is most commonly seen in myopathic conditions that result in a loss of muscle fibers as well as some NMJ disorders. This loss causes less force to be generated per motor unit. Thus, more motor units must now be called upon.
- **Reduced recruitment (neuropathic):**
  - Refers to the firing of fewer than expected MUAPs with even a maximal contraction.
  - It is most commonly seen in neuropathic conditions but can also be in severe myopathies.

Recruitment Parameters

- **Firing Rate (FR) (Figure 5–70):**
  - This is the number of times a MUAP fires per second. It is expressed in Hz and is calculated by dividing 1,000 by the interspike interval (II) measured in ms. FR = 1,000/II.

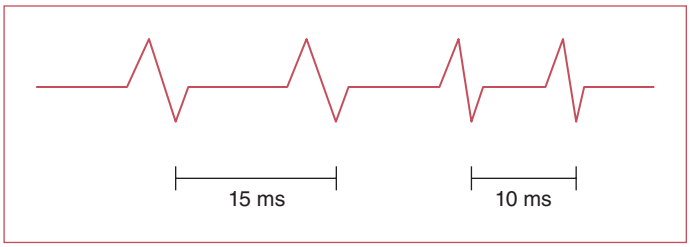


FIGURE 5–70 Firing rate: 1,000/13 = approximately 75.

- **Recruitment frequency (RF):**
  - This is the FR of the first MUAP when a second MUAP begins to fire. It is initiated by an increase in the force of a contraction. Normal is considered 20 Hz or below. Values above this correlate with a neuropathic process.
- **Recruitment interval (RI):**
  - This is the interspike interval (in milliseconds) between two discharges of the same MUAP when a second MUAP begins to fire. It is initiated by an increase in the force of a contraction. Normal is considered approximately 100 ms.
- **RI Versus RF:**
  - The following parameters help distinguish between a neuropathic and myopathic process:

FIRING PATTERN	NEUROPATHY	MYOPATHY
Recruitment interval	↓	↑
Recruitment frequency	↑	↓

  - **Decreased RI (Increased RF):**
    - A loss of motor units restricts additional motor unit activation to increase contractile force. This causes the first motor unit to fire more rapidly until a second motor unit finally joins in. This shortens the interval between successive MUAPs from one motor unit.
  - **Increased RI (Decreased RF):**
    - A loss of muscle fibers causes a second motor unit to join in early to help increase contractile force. This occurs before the first motor unit has the opportunity to increase its firing frequency. This lengthens the interval between successive MUAPs from one motor unit.
  - **Recruitment ratio (RR):**
    - This has been used to represent recruitment capabilities, especially when a patient demonstrates difficulty in controlling a contractile force. It is calculated by dividing the FR of the first MUAP by the number of different MUAPs on the screen. A motor unit firing at 10 Hz when two different MUAPs are viewed on the screen demonstrates an RR of 5. 10 Hz/2 different MUAP = 5. The normal RR is considered <10 (**Figure 5–71**).

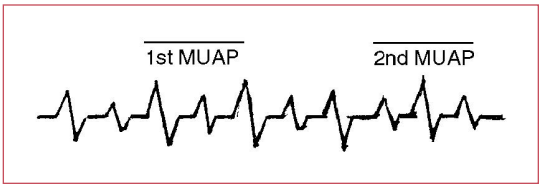


FIGURE 5–71 Recruitment ratio: 10 Hz/2 different MUAP = 5. MUAP, motor unit action potential.

INTERFERENCE PATTERN (FIGURE 5–72)

- This is a qualitative or quantitative description of the sequential appearance of MUAPs.
- It is the electrical activity recorded from a muscle during a maximum voluntary contraction. It is composed of *recruitment* plus *activation*.
- Activation is the ability of a motor unit to fire faster to produce a greater contractile force and is controlled by a central process.
- It can be decreased in CNS diseases, pain, and hysteria.

- Furthermore, if a patient is asked to generate a force and only a few MUAPs are seen while the frequency (Hz) continues to remain low, it can indicate decreased activation from poor patient cooperation and is not the result of abnormal recruitment.

PATTERNS (FIGURE 5–72)	PRESENTATION
Complete	No individual MUAPs can be seen. A full screen represents four to five MUAPs.
Reduced	Some MUAPs are identified on the screen during a full contraction.
Discrete	Each MUAP can be identified on the screen during a full contraction.
Single unit	One MUAP is identified on the screen during a full contraction.

MUAP, motor unit action potential.

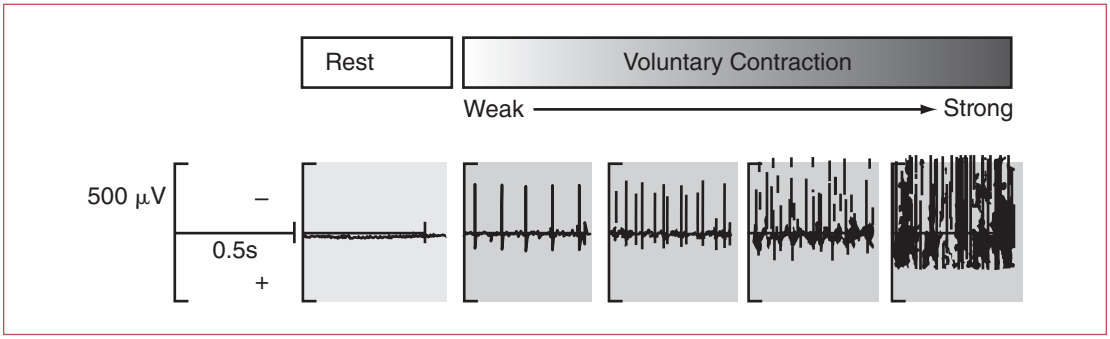


FIGURE 5–72 Interference patterns: Representation of normal recruitment patterns.

RADICULOPATHY

GENERAL (FIGURE 5–73)

- This is a pathologic process affecting the nerves at the root level. In descending order, it most commonly presents as pure sensory complaints, sensorimotor complaints, or pure motor complaints. This involves multiples, as each dermatome is innervated by one spinal nerve root due to the larger size of the sensory fibers, rendering them more prone to injury.
- NCS are typically normal. A pure sensory injury would demonstrate a negative EMG, and sensory NCS would be normal due to sparing of the DRG.
- Although highly unusual, it can occur without structural abnormalities on diagnostic imaging studies (e.g., MRI) or EDX abnormalities on EMG.

ETIOLOGY

- **Common:**
  - *Disc herniation*: Most common; typically seen in adults <50 years of age
  - *Spinal stenosis*: Typically seen in adults >50 years of age
- **Uncommon: "HI MADAM":**
  H—Herpes zoster
  I—Inflammatory: Tuberculosis (TB), Lyme disease, HIV, syphilis, cryptococcus, and sarcoidosis
  M—Metastasis
  A—Arachnoiditis: Myelogram, surgery, steroids, and anesthesia
  D—Diabetes mellitus
  A—Abscess
  M—Mass in the spine: Meningioma, neurofibroma, leukemia, lipoma, synovial cysts, and hematoma

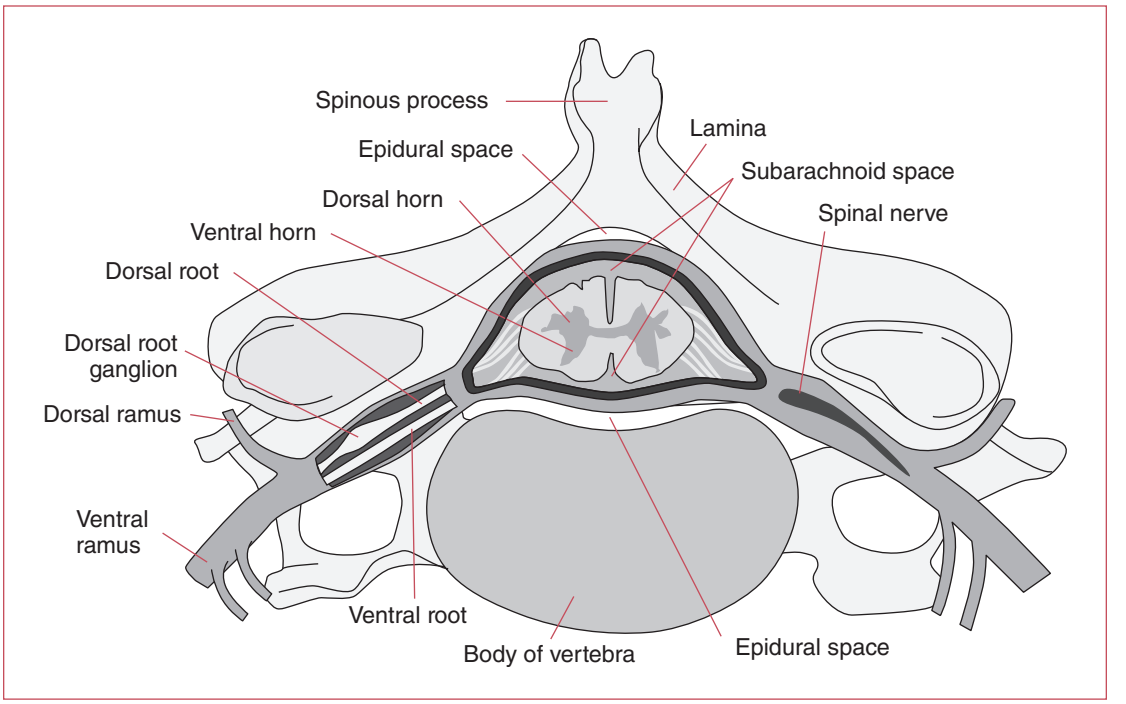


FIGURE 5–73 Cervical spine cross section.

CLINICAL FINDINGS (TABLE 5–23)

TABLE 5–23 Clinical Findings of Radiculopathies

NERVE ROOT	REDUCED/ABSENT REFLEX	WEAKNESS	NUMBNESS/PARESTHESIAS
C5	Biceps brachii	Elbow flexion	Lateral shoulder
C6	Brachioradialis	Elbow flexion	Radial forearm, thumb, index finger
C7	Triceps brachii	Elbow extension	Middle finger
C8	None	Finger flexion	Ring, little finger, hypothenar eminence
T1	None	Finger adduction	Ulnar forearm
L1	None	Hip flexion	Inguinal region
L2	None	Hip flexion/knee extension	Inguinal region/proximal anterolateral thigh
L3	Patellar tendon	Hip flexion/knee extension	Inguinal region/mid-anterolateral thigh
L4	Patellar tendon	Knee extension Ankle dorsiflexion	Anterolateral thigh/anteromedial calf/medial foot
L5	Medial hamstring	Ankle dorsiflexion Hallux extension Foot eversion	Posterolateral thigh/calf and dorsal foot
S1	Achilles tendon	Plantar flexion Foot eversion	Posterior thigh/calf and lateral toes and heel

ELECTRODIAGNOSTIC FINDINGS

- The guidelines for evaluation of a radiculopathy should include at least one motor and one sensory NCS, as well as needle EMG examination including a sufficient number of muscles representing all the relevant myotomes.
- **NCS:**
  - The NCS portion of EDX studies are rarely abnormal in cases of radiculopathies. The primary role of NCS is to evaluate if there are superimposed neurologic pathologies to explain the patient's symptoms.
  - SNAP: Normal if the lesion is located proximal to the DRG
  - CMAP: Typically normal due to multilevel spinal nerve root innervation of a muscle. Reduced amplitudes may be seen in cases with more severe axonal lesions affecting the main spinal nerve root innervating a muscle. The lesion is distal to the motor neuron cell body. It can be normal if the injury is purely demyelinating, incomplete, or reinnervation has occurred.
- **Late responses:**
  - H-reflex: Possibly abnormal in an S1 radiculopathy but not pathognomonic.
  - F-waves: Not sensitive or specific for a radiculopathy. Muscles have more than one root innervation, which can result in a normal latency.
- **Needle EMG:**
  - Needle EMG criteria for diagnosing a radiculopathy includes abnormal findings in two or more muscles innervated by the same spinal nerve root but by different peripheral nerves. Ideally, six muscles (five peripheral muscles + paraspinals) should be evaluated (Dillingham, 2013).
  - 🕮 The optimal number of muscles to screen for a cervical or lumbar radiculopathy is six (five peripheral muscles + paraspinal muscles). If one of the six muscles is abnormal, then further muscles should be evaluated. A priority is placed on including weak muscles in the evaluation.
  - 🕮 Classically, FIBs or PSWs should be found in two different muscles innervated by two different peripheral nerves originating from the same root. They may not be found if the lesion is demyelinating neuropathies, pure sensory nerve injuries, chronic nerve injuries, or missed by random sampling.
- **SSEP:**
  - *Advantage*: It monitors sensory pathways and proximal demyelinating injuries.
  - *Disadvantage*: The long pathway monitored can mask focal lesions between the recording sites and is not considered useful for radiculopathies.

🕮 CERVICAL MYOTOMES (TABLE 5–24)

TABLE 5–24 Cervical Myotomes Affected Secondary to HNP

ROOT	C3/C4	C5	C6	C7	C8
Etiology	C2–C3 + C3–C4 HNP	C4–C5 HNP	C5–C6 HNP	C6–C7 HNP	C7–T1 HNP
Muscles involved	Clinical diagnosis No discrete myotomal patterns Innervates the posterior and lateral scalp Patient may complain of headaches C2 and C3 nerves become the greater and lesser occipital nerve, respectively	Rhomboids Deltoid Biceps Supraspinatus Infraspinatus Brachialis BR Supinator Paraspinals	Deltoid Biceps BR Supraspinatus Infraspinatus Supinator PT FCR Paraspinals	PT FCR EDC Triceps Paraspinals	Triceps FCU FDP Abductor digiti minimi First dorsal interossei PQ Abductor pollicis brevis Paraspinals

BR, brachioradialis; EDC, extensor digitorum communis; FCR, flexor carpi radialis; FCU, flexor carpi ulnaris; FDP, flexor digitorum profundus; HNP, herniated nucleus pulposus; PQ, pronator quadratus; PT, pronator teres.

LUMBOSACRAL MYOTOMES (TABLE 5–25)

TABLE 5–25 Lumbar Myotomes Affected Secondary to HNP

ROOTS	L2/L3/L4	L5	S1	S2/S3/S4
Etiology	L1–L2/L2–L3/L3–L4 HNP	Posterolateral L4–L5 HNP	Posterolateral L5–S1 HNP	Iatrogenic, cauda equina, spinal stenosis
Muscles involved	Iliopsoas Iliacus Gracilis Adductor longus Vastus medialis TA Paraspinals Difficult to distinguish between an L2 and L3 radiculopathy	Gluteus maximus Gluteus medius TFL TA Medial gastrocnemius Medial hamstring TP Peroneus (fibularis) longus Paraspinals	Gluteus maximus Gluteus medius TFL Medial gastrocnemius Medial hamstring Peroneus (fibularis) longus TP Paraspinals	Abductor hallucis Abductor digiti quinti Needle exam of the external anal sphincter Other clinical presentations to monitor: Bulbocavernosus reflex, anal wink, external sphincter tone, and bowel and bladder function

HNP, herniated nucleus pulposus; TA, tibialis anterior; TFL, tensor fascia lata; TP, tibialis posterior.

MUSCLES WITH DUAL INNERVATION (TABLE 5–26)

TABLE 5–26 Muscles With Dual Innervation

MUSCLE	NERVES	
Pectoralis major	Medial pectoral	Lateral pectoral nerve
Brachialis	Musculocutaneous nerve	Radial nerve
Flexor digitorum profundus	Median nerve (AIN)—FDP 1,2	Ulnar nerve—FDP 3,4
Lumbricals	Median nerve	Ulnar nerve
Flexor pollicis brevis	Median nerve	Ulnar nerve
Pectineus	Femoral nerve	Obturator nerve
Adductor magnus	Sciatic nerve (tibial portion)	Obturator nerve
Biceps femoris	Sciatic nerve (tibial portion)	Sciatic nerve (peroneal [fibular] portion)

AIN, anterior interosseus nerve; FDP, flexor digitorum profundus.

CHRONOLOGY OF ELECTRODIAGNOSTIC FINDINGS (TABLE 5–27)

TABLE 5–27 Chronology of Electrodiagnostic Findings

TIME	ABNORMALITY
0	Decreased recruitment Decreased recruitment interval Prolonged F-wave Abnormal H-reflex (S1 radiculopathy)
4 days	Decreased compound motor action potential amplitude (approximately 50% compared with opposite side) in severe cases
1 week	Abnormal spontaneous activity occurs first in the paraspinals They can be normal if: • They become reinnervated • The posterior primary rami are spared They can be the only abnormal finding 10%–30% of the time

(*continued*)

TABLE 5–27 Chronology of Electrodiagnostic Findings (*continued*)

TIME	ABNORMALITY
2 weeks	Abnormal spontaneous activity beginning in the limbs
3 weeks	Abnormal activity present in both the paraspinals and limbs
5–6 weeks	Reinnervation begins
6 months–1 year	Increased amplitude from reinnervated motor unit. Reinnervation complete.
Every 3–4 months	Serial EMG can be performed in intervals to monitor for reinnervation as clinically warranted.

EMG, electromyography.

PLEXOPATHIES

GENERAL

- This is a pathologic process typically occurring distal to the DRG and proximal to the peripheral nerves. Abnormalities can appear diffuse and will not follow any particular dermatomal or myotomal distribution.

ETIOLOGY

- Common etiologies include:
  - Trauma: Traction, transection, obstetrical injuries, compression, and hemorrhage
  - Cancer (tumor and radiation therapy)
  - Idiopathic (neuralgic amyotrophy)

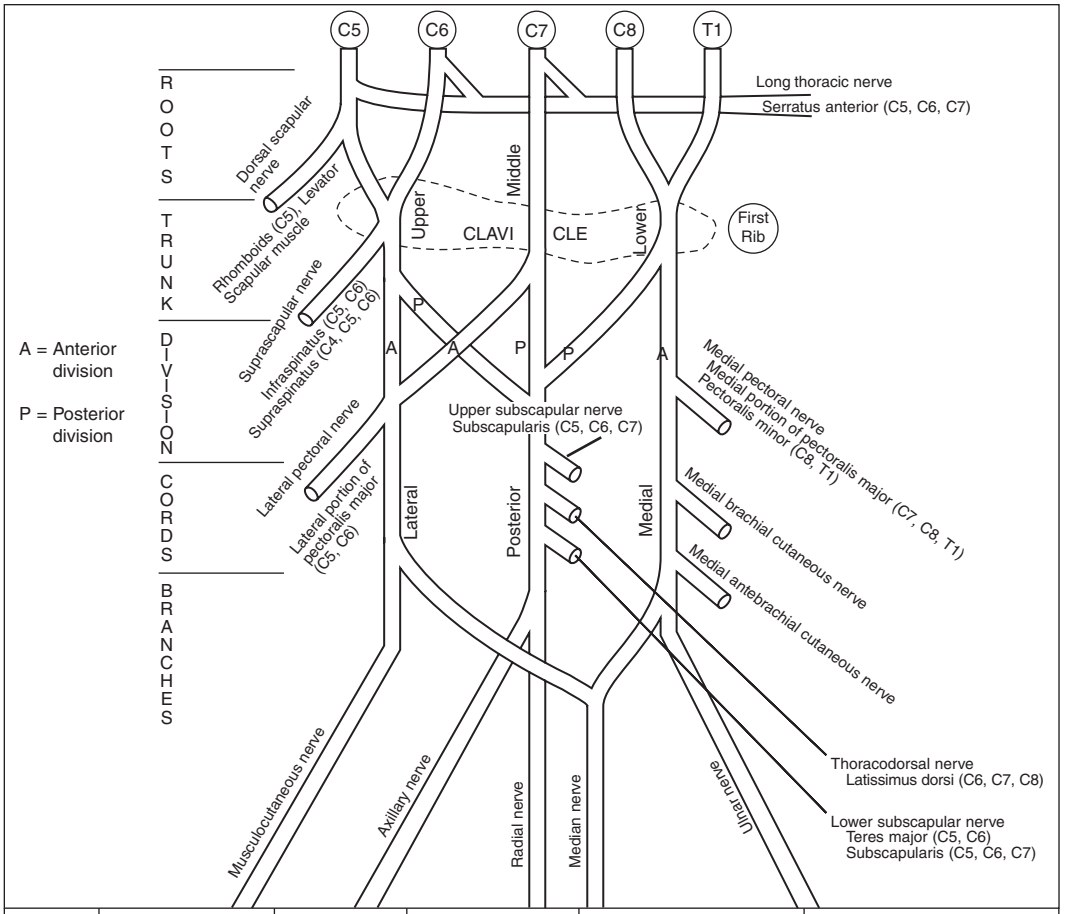
ELECTRODIAGNOSTIC FINDINGS

- Active recruitment of the weakened muscles helps to rule out more involved processes, such as neurotmesis. However, a complete neuropraxic block can appear identical to neurotmesis or axonotmesis in an acute injury if active recruitment cannot be generated.
- **NCS:**
  - Abnormal SNAP and CMAP patterns correspond to the site of plexus injury.
  - The SNAP helps to localize if a lesion is proximal or distal to the DRG. Decreased SNAP amplitudes result from axon loss distal to the DRG. If the SNAP is preserved, but there is marked abnormality to the CMAP, then the injury is proximal.
  - The distal CMAP amplitude is the main prognostic factor in a plexopathy because it represents axonal loss in such conditions, and side-to-side comparison should be performed.
  - See specific injury patterns in this section for more details.
- **Late responses:**
  - **F-waves** may be delayed or absent in plexopathies but are nonspecific, as they cannot localize a focal lesion.
  - **H-reflex:** Helpful to evaluate the S1 pathway but not pathognomonic
- **Needle EMG findings:**
  - Abnormal activity in the peripheral muscles in the distribution of plexus injury (e.g., upper trunk distribution) but with normal paraspinal activity

THE BRACHIAL PLEXUS

Anatomy (Figure 5–74)

- **Origin:**
  - Nerve fibers originate from the ventral rami of the C5–T1 nerve roots, which further divide to form the trunks, division, cords, and terminal nerve branches.



Nerves	Musculocutaneous	Axillary	Radial Nerve	Median Nerve	Ulnar Nerve
C5 C6	• Biceps • Brachialis	• Deltoid • Teres minor	• Supinator		
C5 C6 C7	• Coracobrachialis		• Brachioradialis		
C6 C7				• Pronator teres • Flexor carpi radialis	
C6 C7 C8			• ECR longus • Triceps		
C7 C8			• ECR brevis • Ext. dig. • EIP • EDM • ECU • Abd. poll. longus • Ext. poll. brevis • Ext. poll. longus	• Palmaris longus	• Flexor carpi ulnaris
C7 C8 T1			• Anconeus	• FDS—4 muscles	
C8 T1				• FDP—2 muscles • FPL • Pronator quad • Lumbricals—2 muscles • Opponens pollicis • Abductor pollicis brevis • Flex poll. brevis 1/2	• FDP—2 muscles • Dorsal interossei—4 muscles • Palmar interossei—3 muscles • Lumbricals—2 muscles • Add poll.—1 muscle • Flex poll. brevis 1/2 • Hypothenar muscles – Oppon. dig. min. – Abd. dig. min. – Flex. dig. min. • Palmaris brevis

FIGURE 5–74 Brachial plexus. (Root levels of individual muscles are identified to the left of the table.) ECR, extensor carpi radialis; ECU, extensor carpi ulnaris; EDM, extensor digiti minimi; EIP, extensor indicis proprius; FDP, flexor digitorum profundus; FDS, flexor digitorum superficialis; FPL, flexor pollicis longus.

- **Course:**
  - The ventral rami emerge between the anterior and middle scalene muscles. In the posterior triangle of the neck, C5 and C6 form the upper trunk, C7 forms the middle trunk, and C8 and T1 form the lower trunk.
  - The trunks pass the clavicle and form anterior and posterior divisions to become cords. The cords are named in their relation to the axillary artery.
  - The three posterior divisions of the upper, middle, and lower trunk form the posterior cord. The lateral cord is formed from the anterior divisions of the upper and middle trunk, and the medial cord is formed by the anterior divisions of the lower trunk.
  - The lateral cord splits to form the musculocutaneous branch and also fuses with the medial cord to form the median branch. The posterior cord splits into the radial and axillary branches, and the medial cord splits to contribute to the median branch and the ulnar branch.

BRACHIAL PLEXUS INJURIES

Upper Trunk Brachial Plexopathy/Erb's Palsy/"Stinger"

- *General*: This injury involves the C5–C6 nerve roots of the upper trunk.
- *Etiology*: It can occur from nerve traction or compression or from an obstetrical injury. It can be sports-related (stinger) and involve the C5–C6 nerve roots or the upper trunk.
- *Clinical presentation*:
  - A classic manifestation is the *waiter's tip* position. The arm becomes adducted (deltoid and supraspinatus weakness), internally rotated (teres minor and infraspinatus weakness), extended (bicep and brachioradialis [BR] weakness), pronated (supinator and BR weakness), with the wrist flexed (extensor carpi radialis longus and brevis weakness; **Figure 5–75**).
- *EDX pearl*:
  - Stimulate at the tip of the C6 transverse process over the trunks of the brachial plexus to assess Erb's point (**Figure 5–76**)
- *Treatment*: Rehabilitation, intermittent splinting, and activity restriction

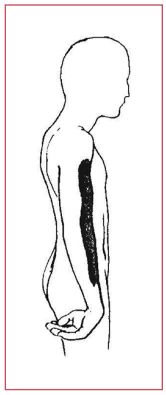


FIGURE 5–75 Waiter's tip position: The area of skin that is usually anesthetic is shaded.

Lower Trunk Brachial Plexopathy/Klumpke's Palsy

- *General*: This injury involves the C8–T1 nerve roots or lower trunk.
- *Etiology*: It can occur from an obstetrical traction injury (Klumpke's palsy), forced adduction seen in a motor vehicle accident (MVA), falls, shoulder dislocations, Pancoast tumor, thoracic outlet syndrome (TOS), and so on.
- *Clinical presentation*:
  - The patient may have wasting of the small hand muscles and a claw hand deformity (lumbrical weakness; **Figure 5–77**). The shoulder girdle muscle function is preserved.
- *EDX pearl*:
  - The preservation of a SNAP potential may indicate a nerve root avulsion. Avulsions may be associated with this location of injury due to the lack of protective support at the C8 and T1 roots. In addition, medial antebrachial cutaneous sensory response will be absent or reduced.
  - *Treatment*: Rehabilitation with incomplete lesions or surgical exploration with a nerve root avulsion injury

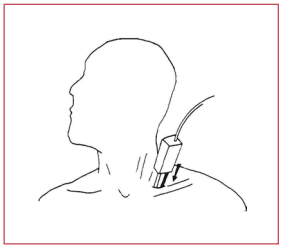


FIGURE 5–76 Erb's point stimulation.

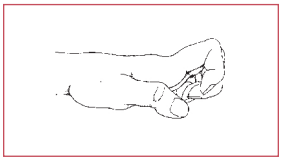


FIGURE 5–77 Ulnar claw hand.

Thoracic Outlet Syndrome (Figure 5–78; Lower Trunk Plexopathy)

- TOS (**Figure 5–78**) is a type of lower trunk plexopathy due to vascular or neurogenic causes.
- **Vascular TOS:**
  - *General*: This injury involves the subclavian artery, subclavian vein, or axillary vein.

– *Etiology*: It can occur from a pathology resulting in arterial or venous compromise.
– *Clinical presentation.*
  ■ Arterial involvement may manifest as limb ischemia, necrosis, vague pain, or fatigue, with decreased color and temperature. Venous involvement can manifest as a bluish, swollen, and achy limb.

• **Neurogenic TOS:**
– *General*: This injury is rare and may actually be seen in only 1 of 1,000,000 patients.
– *Etiology* (**Figure 5–78**):
  ■ It can occur from compression of the lower trunk of the brachial plexus between a fibrous band, between the first cervical rib and clavicle (costoclavicular syndrome), muscular entrapment by the scalenes (anterior and middle scalene syndromes), or pectoralis minor muscle (pectoralis minor syndrome).

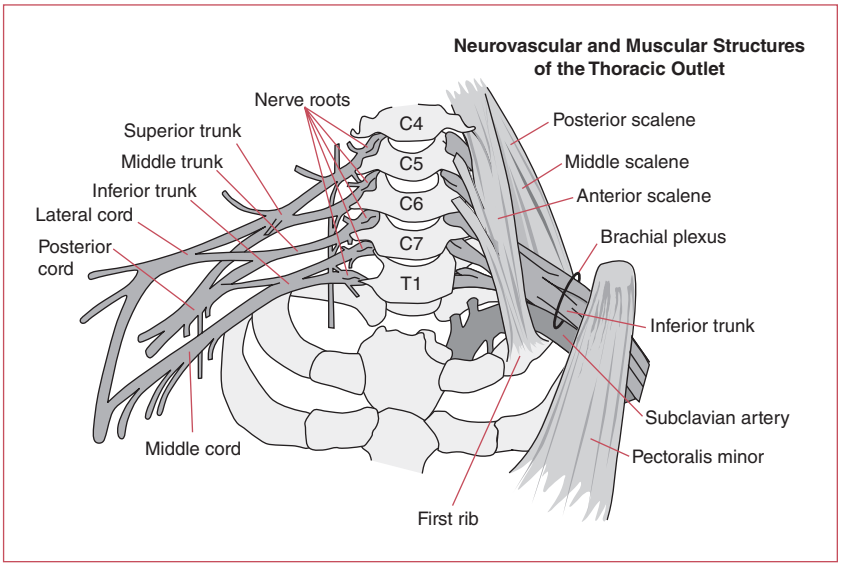


FIGURE 5–78 The thoracic outlet.

– *Clinical presentation*:
  ■ The patient may have pain and numbness along the medial aspect of the forearm and hand, which increases with overhead activity. Discomfort can also be noted in the neck, clavicle, and axilla.
  ■ Hand muscle wasting may also be noted (median thenar > ulnar intrinsics).
  ■ A maneuver to assess the neurovascular bundle is Adson's test (**Figure 5–79**). This is performed by abduction, extending and externally rotating the patient's arm. While monitoring the radial pulse, have the patient rotate the head toward the arm (the side of the lesion). A decrease or loss of pulse may be related to a compression of the subclavian artery, indicating compromise to the complex.
– *EDX pearl*:
  ■ 🕮 Abnormal findings that can be noted are decreased amplitudes for the median CMAP, ulnar SNAP/CMAP, and medial antebrachial cutaneous studies. The median SNAP is spared. Abnormal spontaneous activity can also occur in the median and ulnar hand muscles (lower trunk) on EMG.

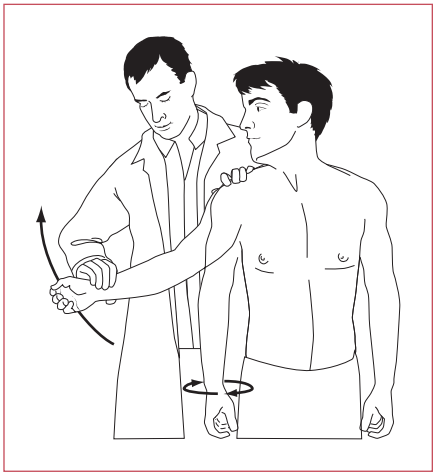


FIGURE 5–79 Adson's test.

- *Treatment*:
  - Rehabilitation with a focus on range-of-motion (ROM) exercises, stretching of appropriate muscles (anterior/middle scalenes, pectoralis minor, trapezius and levator scapulae), strengthening of the scapular stabilizers (upper/middle trapezius and rhomboids), and focus on postural mechanics to address possible entrapment syndromes against the first rib.
  - Surgery can be indicated for a first rib or fibrous band resection.

Neuralgic Amyotrophy

- *General*: This injury can include various nerves of the brachial plexus.
  - Other names include Parsonage–Turner syndrome, brachial neuritis, brachial neuropathy, idiopathic brachial plexopathy, shoulder-girdle neuritis, and paralytic brachial neuritis.
- *Etiology*:
  - Idiopathic, inflammatory, and immune-mediated process
- *Clinical presentation*:
  - Individuals of all ages may be affected, and there is a male predominance. The most common initial symptoms are pain of abrupt onset, often severe, usually in the shoulder or periscapular region. It can be exacerbated by abduction and rotation.
  - The pain may resolve in hours to days but typically begins to improve in 2 to 3 weeks, in association with the development of weakness in a patchy fashion.
  - Bilateral involvement occurs in about one-third of patients, usually asymmetrically. The patchy or multifocal involvement is common and is a hallmark of this syndrome.
- *EDX pearl*:
  - Abnormal motor and sensory NCS and/or spontaneous activity can occur in a variety of combinations: Mononeuropathy (suprascapular nerve, long thoracic nerve, axillary nerve, anterior interosseus nerve [AIN], spinal accessory nerve) or plexopathy. Muscles innervated by the C5 and C6 levels are more commonly affected.
- *Treatment*:
  - Interventions may include rehabilitation to prevent contractures. Neuralgic amyotrophy can resolve spontaneously.

Neoplastic Versus Radiation Plexopathy (Table 5–28)

- *General*:
  - Brachial plexopathies can be caused by compression from tumors and/or from their treatments.
  - A primary plexus tumor can arise from schwannomas, neuromas, and neurofibromas.
  - Secondary plexus involvement can arise from a Pancoast tumor from the lung or breast.
  - Radiation therapy can cause neural fibrosis and constriction of the vasa nervorum, leading to destruction of the axon and Schwann cells. This can occur months or years after radiation treatment.
- *Clinical presentation/EDX pearl*:

TABLE 5–28 Neoplastic Versus Radiation Plexopathy

CHARACTERISTICS	RADIATION	TUMOR
Common site of injury	Upper trunk	Lower trunk
Clinical presentations	Myokymia on EMG	Horner's syndrome
Sensation	Painless	Painful

EMG, electromyography.

Nerve Root Avulsion (Figure 5–80)

- *General*:
  - A severe plexus injury can lead to disruption of the nerve roots. This can occur from a traction injury that disrupts the protective connective tissue support. The C8 and T1 roots have less protection and are the most common site of tearing from spinal cord attachments. MRI is valuable in differentiating between a nerve root stretch and an avulsion.
- *Clinical presentation*:
  - The patient complains of absent sensation or muscle contraction from the muscles innervated by the roots involved. This may manifest as a flail shoulder.

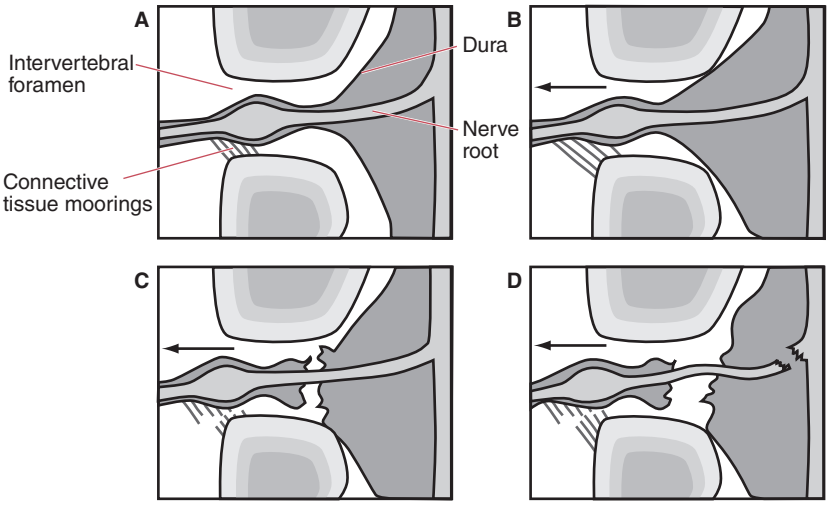


FIGURE 5–80 Nerve root avulsion. (A) Normal nerve root exiting the intervertebral foramen. (B) Slight traction with dura plugging the foramen and support by tissue moorings. (C) Ruptured dura and moorings. (D) Ruptured roots. *Note:* Arrows depict direction of traction forces.

- *EDX pearl*:
  - 🕮 Absent CMAPs with normal SNAPs. Needle EMG reveals absent recruitment and abnormal spontaneous activity in a myotomal distribution of the avulsed nerve root, including the paraspinals.

LUMBOSACRAL PLEXUS (FIGURE 5–81)

Anatomy

- **Origin:**
  - Nerve fibers originating from the ventral rami of L1, L2, L3, and L4 roots form the *lumbar plexus*.
  - Nerve fibers originating from the ventral rami of L4, L5, S1, S2, S3, and S4 roots form the *sacral plexus*.
- **Course:**
  - The ventral rami then divide to form anterior and posterior divisions in each plexus.
  - The anterior division of the lumbar plexus forms the obturator nerve, while the posterior division forms the femoral nerve and the lateral cutaneous nerve of the thigh (lateral femoral cutaneous nerve).

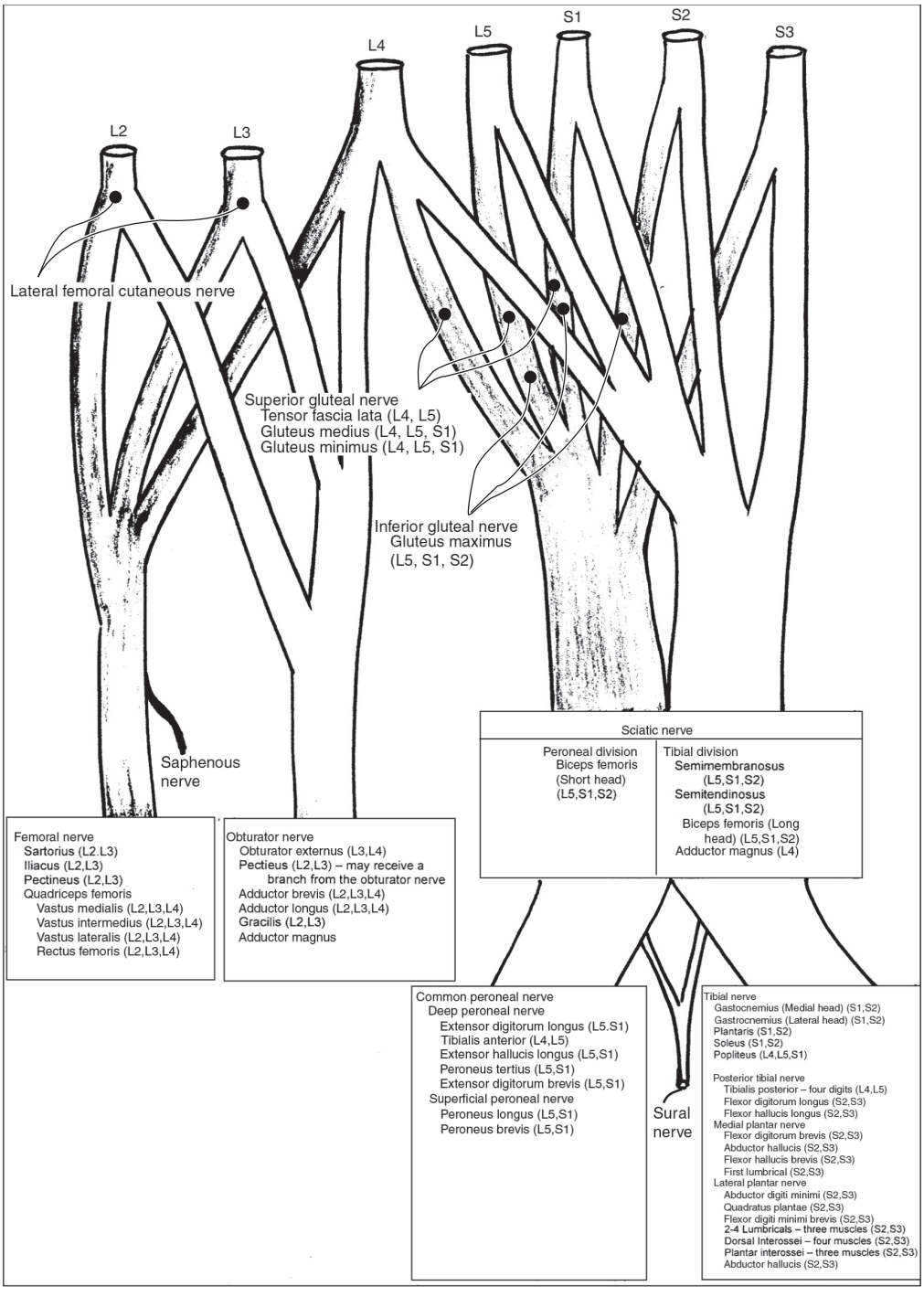


FIGURE 5–81 The lumbosacral plexus and innervations.
Note: The terms peroneus and fibularis are used interchangeably. The words "peroneus" and "peroneal" in the figure have been kept; however, please note that peroneus = fibularis; peroneal = fibular.

- Terminal branches directly off the lumbar plexus include:
  - Iliohypogastric nerve
  - Ilioinguinal nerve
  - Genitofemoral nerve
- In the sacral plexus, the tibial portion originates from the anterior division, while the common peroneal (fibular) nerve originates from the posterior division. The lumbosacral trunk forms from the L4 and L5 nerve fibers connecting the lumbar to the sacral plexus and travels over the pelvic brim.
- Terminal branches directly off the sacral plexus include:
  - Superior gluteal nerve
  - Inferior gluteal nerve

LUMBOSACRAL PLEXOPATHIES

- Multiple potential etiologies:
  - Neuralgic amyotrophy: Similar to brachial plexus pathology
  - Neoplastic versus radiation plexopathy: Similar to brachial plexus pathology
  - Retroperitoneal bleed: This can involve a hematoma formation in the psoas muscle.
  - Hip dislocation
  - Obstetric injuries/cephalopelvic disproportion: Presents as a postpartum foot drop
- *EDX pearl*:
  - Lumbar plexus evaluation:
    - SNAP: Lateral cutaneous nerve of the thigh (Lateral femoral cutaneous nerve; L2–L3), saphenous nerve (L4)
    - CMAP: Femoral nerve (L2–L4)
    - EMG: Muscles innervated by the femoral nerve, obturator nerve, and the iliopsoas muscle. Normal paraspinals.
  - Sacral plexus evaluation:
    - SNAP: Superficial peroneal (fibular) nerve (L5), sural nerve (S1)
    - CMAP: Deep peroneal (fibular) nerve (L4–S1), tibial nerve (L5–S2)
    - EMG: Muscles innervated by the tibial nerve, peroneal (fibular) nerve, and superior and inferior gluteal nerves. Normal paraspinals.

UPPER LIMB MONONEUROPATHIES

MEDIAN NERVE

Anatomy

- **Origin: (Figure 5–82)**
  - Nerve fibers from the C5–T1 nerve roots contribute to the upper, middle, and lower trunks → medial and lateral cords → median nerve.
- **Course:**

ARM

- The nerve runs medial to the axillary artery. It continues down the humerus and runs under the ligament of Struthers (LOS) at the medial epicondyle of the humerus.

FOREARM

- Innervations and cutaneous branches:
  - Pronator teres (PT)
  - FCR

- Palmaris longus
- Flexor digitorum superficialis (FDS)
- Palmar cutaneous branch
- The AIN branches from the median nerve to innervate (four Ps):
  - Flexor pollicis longus (FPL)
  - Flexor digitorum profundus (FDP 1 and 2)
  - Pronator quadratus (PQ)

HAND

- Through the carpal tunnel, the median nerve innervates the "LOAF" muscles:
  - Lumbricals (1, 2)
  - Opponens pollicis
  - Abductor pollicis brevis (APB)
  - Flexor pollicis brevis (superficial)
  - (Digital cutaneous branches)

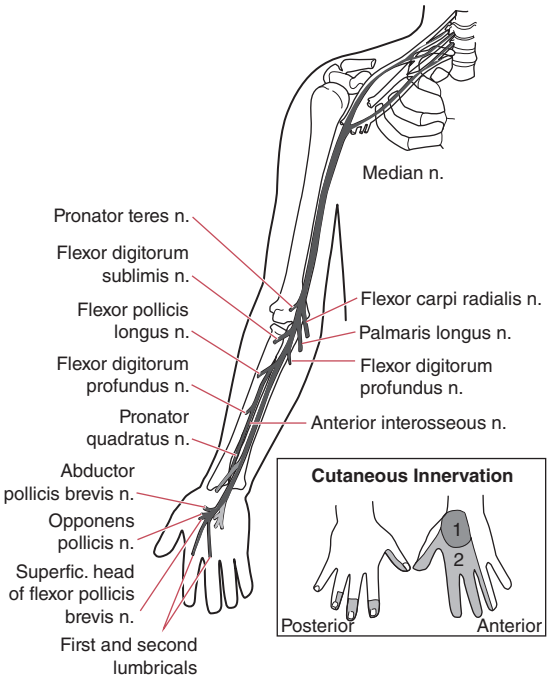


FIGURE 5–82 The median nerve. Cutaneous branches in hand supply: (1) Thenar eminence. (2) Palmar surface as indicated. n., nerve.

Injury

ARM

LIGAMENT OF STRUTHERS (FIGURE 5–83)

- *General*:
  - There is a 2-cm bone spur 3 to 6 cm proximal to the medial epicondyle. It is connected to the epicondyle by a ligament in 1% of the population.
- *Etiology*:
  - The nerve becomes entrapped with the brachial artery under the ligament.
- *Clinical presentation*:
  - The patient may have involvement of *all median nerve innervated muscles*. It can manifest as weakness in grip strength (FDP and FDS weakness) and wrist flexion (FCR weakness). A dull, achy sensation can occur in the distal forearm. There may be difficulty in flexing the second and third digits (FDP weakness) resulting in an active benediction sign. The brachial pulse is possibly diminished.
- *EDX findings*:
  - **NCS:** Abnormal median SNAP and CMAP
  - **EMG:** Abnormal in median nerve-innervated muscles, including the PT
- *Treatment*:
  - Rehabilitation or surgical release

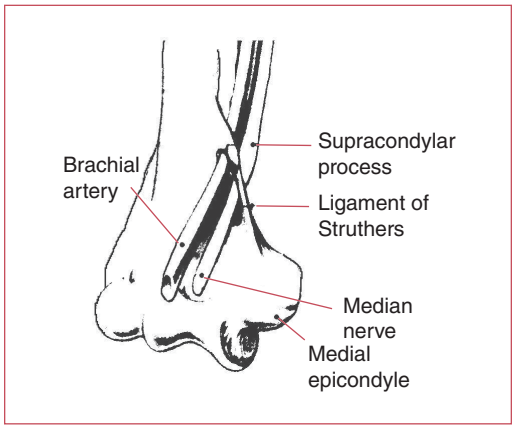


FIGURE 5–83 The ligament of Struthers.

BICIPITAL APONEUROSIS (LACERTUS FIBROSUS; FIGURE 5–84)

- *General*:
  - This is a thickening of the antebrachial fascia attaching the biceps to the ulna. It overlies the median nerve in the proximal forearm.
- *Etiology*:
  - The nerve can be injured by entrapment or hematoma compression resulting from an arterial blood gas or venipuncture.
- *Clinical presentation*:
  - This is similar to the LOS pathology.
- *EDX findings*:
  - This is similar to the LOS pathology.
- *Treatment*:
  - Interventions may include rehabilitation or surgical release.

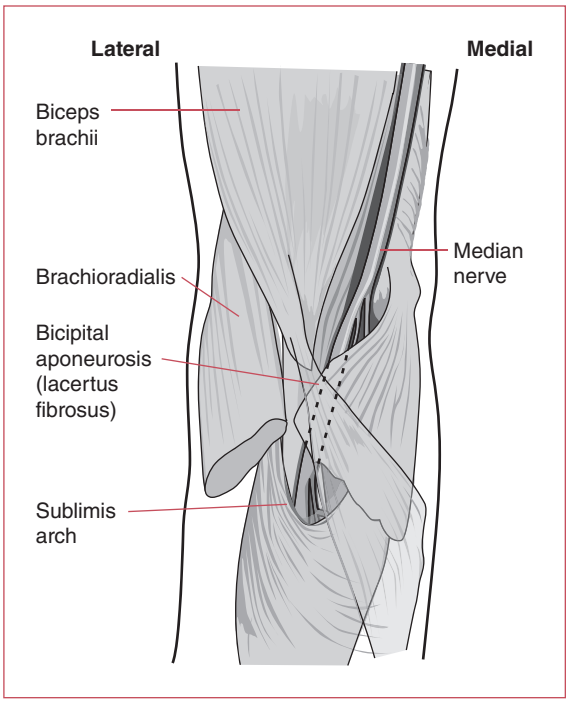


FIGURE 5–84 The lacertus fibrosus.

FOREARM

PRONATOR TERES SYNDROME (FIGURE 5–85)

- *General*:
  - The median nerve passes between the two heads of the PT muscle and continues down under the FDS muscle. An entrapment is commonly named for the muscle compressing the nerve. That muscle is usually spared because it receives its innervation before it is pierced by the nerve. However, the nerve can also contribute to the innervation as it courses through the muscle.
- *Etiology*:
  - The nerve can be injured by compression between the heads of the PT muscle or the bridging fascial band of the FDS muscle.
- *Clinical presentation*:
  - The muscles involved include all median innervated muscles except the PT. The patient may complain of a dull ache of the proximal forearm exacerbated by forceful pronation (PT) or finger flexion (FDS). The forearm and hand muscles may become easily fatigued.
- *EDX findings*:
  - **NCS:** Abnormal median nerve SNAPs and CMAPs
  - **EMG:** Abnormal activity in all median nerve innervated muscles EXCEPT the PT
- *Treatment*:
  - Interventions may include rehabilitation or surgical release.

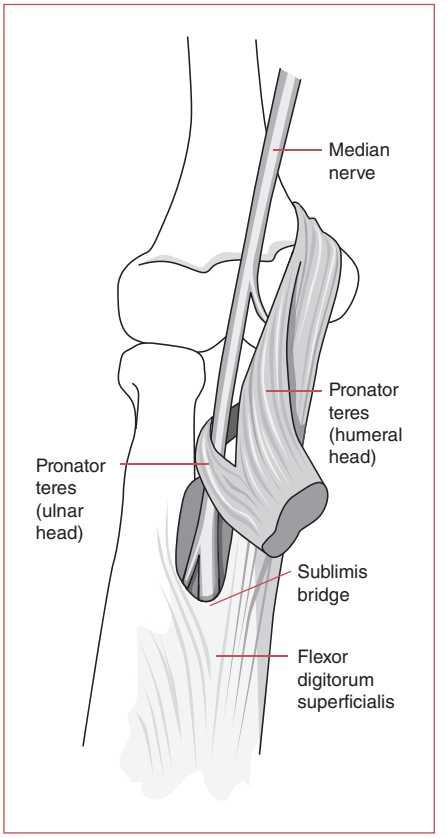


FIGURE 5–85 Median nerve entrapment at the pronator teres or flexor digitorum superficialis (flexor sublimis).

ANTERIOR INTEROSSEUS NERVE SYNDROME

- *General*:
  - This is an injury to a motor nerve branch of the median nerve. It is a pure motor syndrome. It lacks cutaneous innervations but has sensory branches to the wrist joint.

  - 🕮 It supplies the FPL, PQ, and FDP 1, 2 (the four P muscles). The FPL is usually the first muscle affected.
- *Etiology*:
  - The nerve can be injured by an idiopathic process, fracture of the forearm, lacerations, or compression.
- *Clinical presentation*:
  - The patient may demonstrate a positive (abnormal) "OK" sign (**Figure 5–86**) or have difficulty forming a fist (**Figure 5–87**) because of an inability to approximate the thumb and index finger (FPL, FDP weakness). The muscles involved include FPL, PQ, and first and second heads of the FDP (the four P muscles). Sensation is spared.
- *EDX findings*:
  - **NCS:** Normal median nerve SNAP and median nerve CMAP to the APB. Possible abnormal CMAP to the PQ.
  - **EMG:** Abnormal activity in the muscles innervated by the AIN (FPL, FDP 1 and 2, and PQ)
- *Treatment*:
  - Interventions may include rehabilitation or surgical exploration.

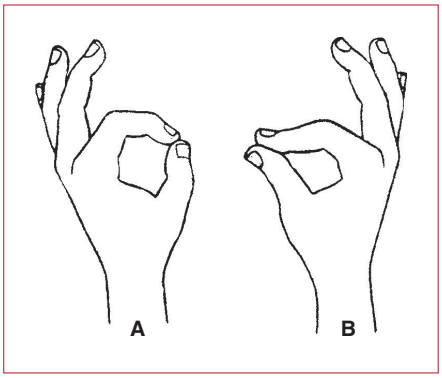


FIGURE 5–86 (A) Normal. (B) Positive OK sign (or pinch sign).

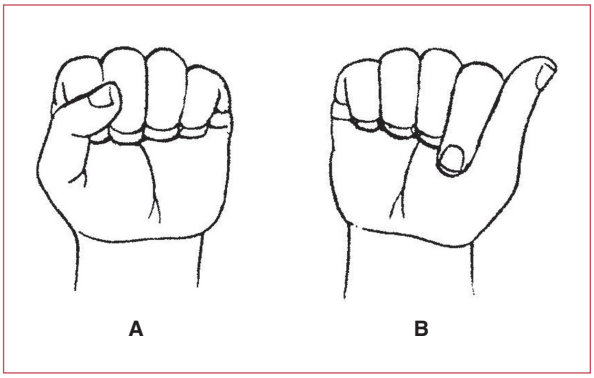


FIGURE 5–87 (A) Normal. (B) Inability to make a fist due to problems incorporating the thumb and index finger.

🕮 *CARPAL TUNNEL SYNDROME*

- *General*:
  - This is a median neuropathy caused by compression of the median nerve in the carpal tunnel and is the most common nerve entrapment in the upper extremity.
  - It is characterized by a clinical syndrome in which nerve damage is coupled with abnormal sensations and pain. It can involve the first, second, third, and 1/2 of the fourth digit.
  - The EDX abnormality alone should not establish the diagnosis, as 10% to 15% of people with clinical carpal tunnel syndrome (CTS) can have a normal study.

CARPAL TUNNEL CONTENTS (FIGURE 5–88)

🕮 CONTENTS: NINE TENDONS AND ONE NERVE	BORDERS
Superficial layer Four FDS tendons One FPL tendon Median nerve (FCR is outside the carpal tunnel)	Transverse carpal ligament Carpal arch bones
Deep layer Four FDP tendons	

FCR, flexor carpi radialis; FDP, flexor digitorum profundus; FDS, flexor digitorum superficialis; FPL, flexor pollicis longus.

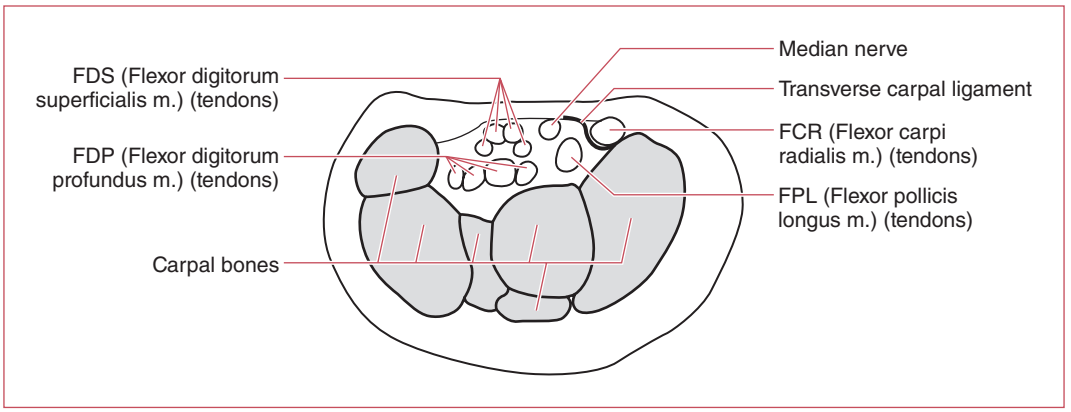


FIGURE 5–88 Cross-section of the carpal tunnel (at the level of the first row of the carpal bones). m, muscle.

- *Etiology*:
  - The nerve can be injured in the carpal tunnel by:
    - Idiopathic process
    - Increased canal volume from thyroid disease, congestive heart failure (CHF), renal failure, mass (tumor, hematoma), and pregnancy (it usually occurs at 6 months and resolves postpartum)
    - Decreased canal volume from a fracture, arthritis, and rheumatoid tenosynovitis
    - Double crush syndrome from diabetes mellitus, cervical radiculopathy, and TOS
- *Clinical presentation*:
  - Symptoms usually begin gradually and are often experienced at night. The thumb, index, and middle fingers are most frequently affected. The most common symptoms of CTS include night numbness, tingling, and pain in the hand; a feeling of electric shock in the fingers or lateral hand.
  - Sensation is abnormal to the lateral 3–1/2 fingers of the hand except at the base of the thumb. Muscle weakness can be noted in "LOAF" muscles (**L**umbricals 1 and 2, **O**pponens pollicis, **A**bductor pollicis brevis, and **F**lexor pollicis brevis).
  - Provocative maneuvers are noted in Table 5–29.
  - The severity of clinical symptoms from CTS does not necessarily correlate with severity of EDX findings, but in general the clinical presentation is as follows:
- ***Mild CTS*:**
  - The patient may complain of numbness, paresthesias, or dysesthesias radiating to the first, second, third, and lateral fourth digits. Symptoms may be exacerbated during sleep and relieved with wrist shaking.
- ***Moderate CTS*:**
  - The patient may complain of continuous sensory deficits in the median nerve distribution, involving the entire palm and radiating proximally. The ability to handle fine objects is impaired.

TABLE 5–29 Provocative Tests for Carpal Tunnel Syndrome

PROVOCATIVE TESTS	DESCRIPTION
Tinel's sign	Percussion of the median nerve at the wrist
Phalen's test	Hold the wrist at 90 degrees flexion for approximately 1 minute
Tourniquet test	Inflated BP cuff reproduction of symptoms at 1 minute
Carpal compression test	Hold thumb compression over the tunnel for 30 seconds
Reverse Phalen's test	Hold the wrist at 90 degrees extension for approximately 1 minute

BP, blood pressure.

- *Severe CTS*:
  - The patient may complain of severe sensory loss and muscular atrophy of the thenar eminence.
- *EDX findings* (Table 5–30):
  - Sensory studies are more sensitive than motor studies because larger sensory fibers are more affected than the slightly smaller motor nerve fibers.
  - Antidromic sensory studies are typically performed and offer the advantage of producing larger amplitudes and are more commonly practiced.

TABLE 5–30 EDX Findings for Carpal Tunnel Syndrome

SEVERITY	NCS	EMG
Mild	SNAP: Prolonged latency CMAP: Normal	Normal
Moderate	SNAP: As above plus decreased amplitude CMAP: Prolonged latency	Normal
Severe	SNAP: Absent CMAP: As above plus decreased amplitude	Abnormal activity

CMAP, compound motor action potential; EDX, electrodiagnostic; EMG, electromyography; NCS, nerve conduction studies; SNAP, sensory nerve action potential.

- *Special studies*:
  - 🕮 On routine median NCS, a demyelinating lesion at the carpal tunnel results in slowing and prolongation of the distal motor and sensory latencies. If the demyelination has resulted in conduction block or secondary axonal loss, the distal CMAP and SNAP amplitudes will be decreased as well. The study of distal segments of the median nerve is helpful in distinguishing CTS from peripheral neuropathy; in CTS, the maximal slowing is across the wrist, whereas in peripheral neuropathy the distal segment is more abnormal.
  - Approximately 10% to 25% of CTS patients have normal routine median conduction studies; in such patients, further testing is performed using more sensitive NCS. Those studies usually involve a comparison of the median nerve to ulnar nerve in the same hand. The common median-versus-ulnar comparison tests are (a) median-versus-ulnar palm-to-wrist mixed-nerve latencies (the typical cutoff is 0.4 ms), and (b) median-versus-ulnar wrist-to-digit-4 sensory latencies (a difference of 0.5 ms is considered abnormal).
- *Treatment*:

Interventions may include the following:

  - *Conservative*
    - Indications: Mild symptoms with no weakness/atrophy or denervation on EMG.
    - Orthotics: Hand splint neutral to 30-degree extension.
    - Medications: Nonsteroidal anti-inflammatory drugs (NSAIDs), or a steroid injection, diuretics, and vitamin B6.
    - Ergonomic modifications
    - Treat underlying medical disorders
  - *Surgical indications*:
    - 🕮 Persistent numbness/paresthesias, severe pain despite a trial of conservative treatment.
    - Severe CTS with severe muscle atrophy typically does not have a high surgical success rate due to the degree of nerve damage that has already occurred.
- *Prognosis*:
  - Poor outcome with conservative management may occur with:
    - Symptoms >10 months in duration
    - Constant paresthesias
    - Positive Phalen's test in <10 seconds

- Weakness, atrophy
- Marked prolonged latency on NCS
- Abnormal spontaneous activity on EMG

ANOMALOUS INNERVATIONS

MARTIN–GRUBER ANASTOMOSIS (FIGURE 5–89)

- *General*:
  - This is a median to ulnar nerve anastomosis. Fibers from the AIN branch of the median nerve anastomose with the ulnar nerve. Alternatively, nerve fibers of the proximal median nerve cross over to the ulnar nerve in the forearm to innervate the adductor pollicis (ADP), abductor digiti minimi (ADM), and most commonly the first dorsal interossei (DI) muscles. This can occur in 15% to 20% of the population.
- 🕮 *EDX findings* (**Figure 5–90**):
  1. 🕮 An initial positive deflection of the median nerve CMAP (**Figure 5–90A**) with median nerve stimulation at the antecubital fossa, which is not seen at the wrist. This is due to the proximal stimulus stimulating the ulnar nerve/muscles through the anastomosis, and the potential from the ulnar muscle fibers reaching the recording electrode via volume conduction before the potential from the median nerve as a result of the median nerve fibers being entrapped and slowed down at the wrist.
  2. 🕮 An increased amplitude of the median nerve CMAP (**Figure 5–90B**) is seen at the elbow compared with the wrist. This is due to simultaneously stimulating the median and the ulnar nerve innervated muscles at the elbow. This can occur without a median nerve entrapment.
  3. An artificially fast CV can be demonstrated. This is a mathematical error due to using a normal

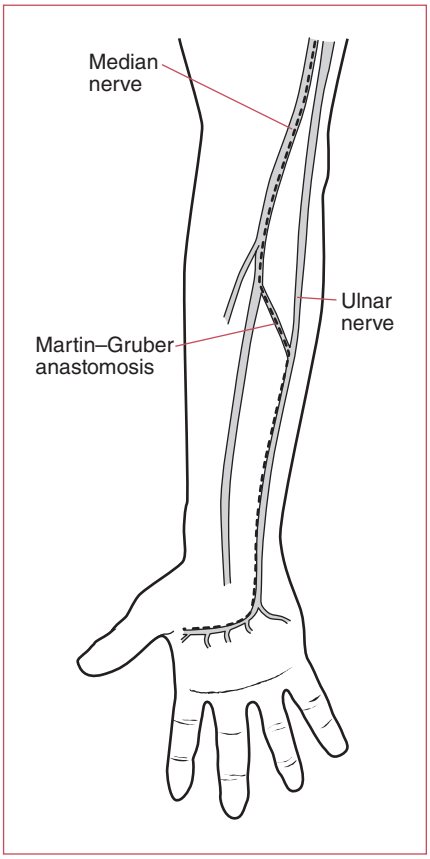


FIGURE 5–89 Martin–Gruber anastomosis.

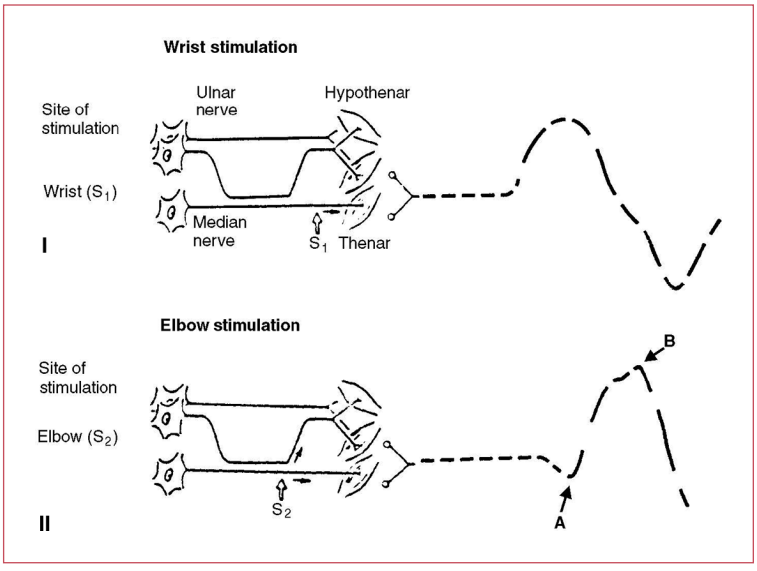


FIGURE 5–90 Martin–Gruber anastomosis with **carpal tunnel syndrome** EDX finding: (A) Initial positive deflection of the CMAP is caused by response of ulnar muscles innervated by the median nerve, which has gotten to the recording electrode earlier, since the aberrant portion of the median nerve is not entrapped. (B) Increased amplitude of the median nerve CMAP is seen at the elbow (as opposed to the wrist). This is due to simultaneously stimulating the median and ulnar innervated muscles at the elbow.
CMAP, compound motor action potential; EDX, electrodiagnostic.

proximal latency reading obtained by stimulating the branch to the ADM. This is subsequently calculated with the prolonged distal latency obtained by the entrapped median nerve, giving a falsely increased CV.

Example
Elbow latency (6.0 ms) – Wrist latency (4.5 ms) = 1.5 ms
Forearm measurement (150 mm)/Forearm latency (1.5 ms) = 100 m/s

📖 *RICHE-CANNIEU ANASTOMOSIS*

- *General*:
  - This is a connection of the recurrent branch of the median nerve in the hand to the deep motor branch of the ulnar nerve. It produces an all ulnar innervated hand.
- *EDX findings*:
  - While recording over the APB muscle, the median nerve CMAP is absent with median nerve stimulation but present with stimulation of the ulnar nerve.

ULNAR NERVE

Anatomy

- **Origin (Figure 5–91):**
  - These nerve fibers originate from the C8–T1 roots. They continue on to contribute to the lower trunk, medial cord, and finally form the ulnar nerve.
- **Course:**

ARM

- The nerve descends along the medial surface of the medial head of the triceps. It runs within a deep groove of thick fascia known as the *arcade of Struthers (AOS)*. It continues posteriorly in a sulcus between the medial epicondyle and olecranon called the *retrocondylar groove*.

FOREARM

- It then continues distally in the cubital tunnel to innervate the:
  - Flexor carpi ulnaris (FCU)
  - FDP (third and fourth)
  - Palmar ulnar cutaneous nerve
  - 📖 Dorsal ulnar cutaneous (DUC) nerve: This branch does not travel through Guyon's canal (arising 5–8 cm more proximally) and is normal in a distal ulnar neuropathy at the wrist but abnormal in more proximal compressions.
  - Dorsal digital nerves

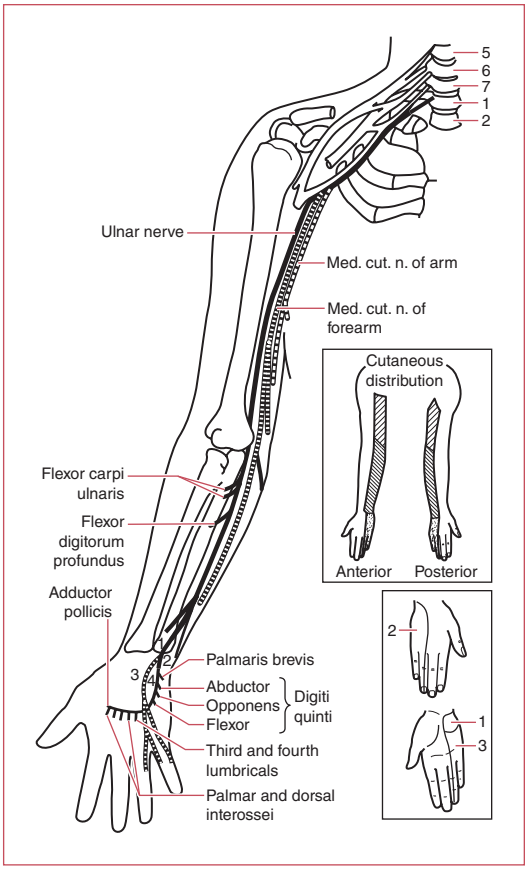


FIGURE 5–91 The ulnar nerve.

HAND

- Through *Guyon's canal*, it splits into three branches:
  - **Superficial sensory branch.**
  - **Hypothenar branch:**
    - Opponens digiti quinti
    - Abductor digiti quinti
    - Flexor digiti quinti

- **Deep motor branch:**
  - Palmaris brevis
  - 4 **D**orsal interossei—("DAB": **Ab**duction)
  - 3 **P**almar interossei—("PAD": **Ad**duction)
  - 2 Lumbricals
  - 1 Adductor pollicis
  - 1/2 Flexor pollicis brevis (deep head)

The 1-1/2 Nerve
- **1-1/2 muscles of the thumb**
- 1 Adductor pollicis
- 1/2 Flexor pollicis brevis (deep head)
- **1-1/2 muscle of the forearm**
- 1 Flexor carpi ulnaris
- 1/2 Flexor digitorum profundus (3, 4)
- **1-1/2 sensation of the fingers**
- 1 Fifth digit
- 1/2 Fourth digit

Injury

ARM

ARCADE OF STRUTHERS (FIGURE 5–92)

- *General*:
  - This is a fascial band in the medial arm that connects the brachialis to the triceps brachii.
- *Etiology*:
  - The ulnar nerve can be injured due to compression under the fascial band.
- *Clinical presentation*:
  - The patient may demonstrate involvement of *all* ulnar nerve innervated muscles.
  - Wrist flexion may result with a radial deviation (FCU weakness).
  - Abnormal sensations may occur in all sensory branches of the ulnar nerve.
  - **Ulnar claw hand (Figure 5–93):** While the hand is at rest, an unopposed pull of the extensor digitorum communis (EDC) causes partial finger flexion of the fourth and fifth proximal interphalangeal (PIP) and distal interphalangeal (DIP) joint due to extension of the metacarpophalangeal (MCP) joint.

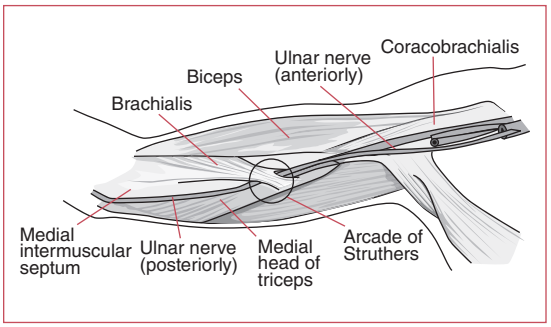


FIGURE 5–92 The Arcade of Struthers.

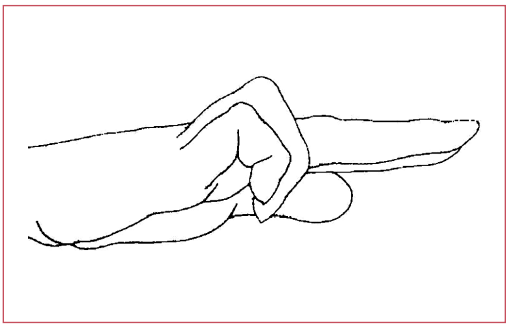


FIGURE 5–93 Ulnar claw hand.

- *Special signs*:
  - ***Froment's sign* (Figure 5–94):** This test demonstrates an inability to hold a piece of paper by the thumb and index finger with pure thumb adduction (ADP weakness). The patient substitutes the median innervated FPL muscle, causing flexion of the interphalangeal joint.
  - ***Wartenberg's sign*:** Inability to adduct the fifth digit (interossei weakness).
- *EDX findings*:
  - **NCS:** Abnormal DUC SNAP; abnormal ulnar SNAP and CMAP
  - **EMG:** Abnormal activity in all the ulnar nerve innervated muscles
- *Treatment*: Interventions may include rehabilitation or surgical release of the AOS.

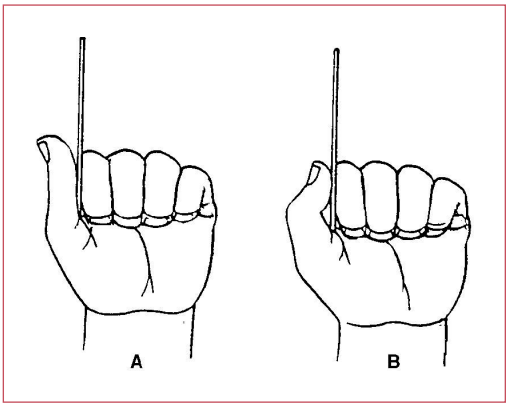


FIGURE 5–94 (A) Normal. (B) Froment's sign.

FOREARM

TARDY ULNAR NERVE PALSY

- *General*:
  - This is an ulnar neuropathy that can occur months to years after a distal humeral fracture.
- *Etiology*:
  - The nerve can be injured secondary to a trauma that results in bone overgrowth or scar formation. Nerve traction can also occur from an increased carrying angle due to a valgus deformity at the elbow.
- *Clinical presentation*:
  - The patient's muscle involvement and complaints are dependent on the site of injury. In most cases, the patient may demonstrate involvement of all the ulnar nerve innervated muscles.
- *EDX findings*:
  - Abnormal ulnar SNAP and CMAP and abnormal EMG of all ulnar muscles
- *Treatment*:
  - Interventions may include rehabilitation or surgical procedure.

CUBITAL TUNNEL SYNDROME

- *General*:
  - This is the most common site of elbow entrapment.
  - It is bordered by the medial epicondyle and olecranon with an overlying aponeurotic band. NCS across the elbow should be performed at 90- to 110-degree elbow flexion (**Figure 5–95**). This will avoid underestimation of actual nerve length, thus causing false positive findings.

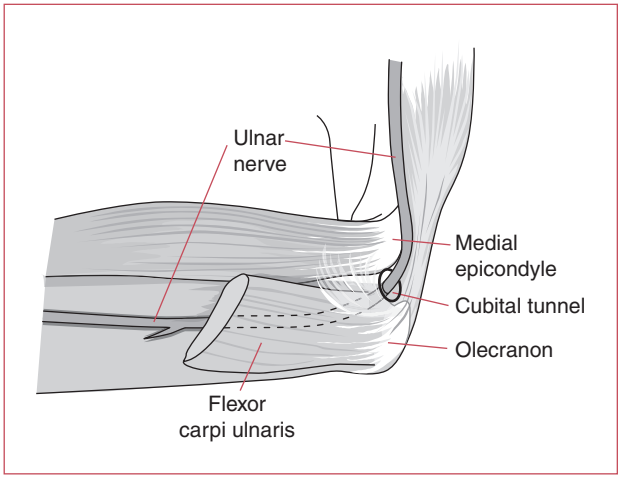


FIGURE 5–95 The cubital tunnel.

- *Etiology*:
  - The nerve can be injured from compression beneath the proximal edge of the FCU aponeurosis or arcuate ligament.
- *Clinical presentation*:
  - The muscles involved include all ulnar nerve innervated muscles with one exception: The FCU *may* or *may not* be involved. The patient may complain of symptoms similar to an injury at the AOS. A positive *Tinel's sign* may demonstrate paresthesias in the distribution of the ulnar nerve with percussion at the ulnar groove.
- *EDX findings*:
  - **NCS:** Motor conduction studies are more useful than sensory NCS. Recording is typically at the ADM. However, the first DI may show earlier abnormalities due to those nerve fascicles being more prone to injury.
  - A decrease in ulnar CMAP amplitude seen both below and above the elbow may be secondary to a Martin–Gruber anastomosis. This can be checked by stimulating the median nerve at the elbow, which will provide a waveform with an amplitude equal to what is considered "missing" from the ulnar nerve stimulation.
    - SNAP: Abnormal ulnar nerve and DUC findings
    - CMAP: Approximately 10 to 15 ms decrease in CV across the elbow (most consistent finding); a 20% drop in amplitude is abnormal
  - **EMG:**
    - Abnormal activity in the ulnar nerve innervated (hand intrinsics > forearm) muscles. Due to nerve fascicle placement in the retrocondylar groove, there is a 70% chance of finding

abnormalities in the hand intrinsics compared with a 25% chance in the ulnar forearm muscles. Furthermore, the FDP has more fascicles that pass through the cubital tunnel and is more reliable to test than the FCU.

- *Treatment*:
  - Interventions may include rehabilitation or surgical release.

HAND

GUYON'S CANAL

- *General*:
  - Different branches of the ulnar nerve can be injured at the wrist (**Figure 5–96**).
- *Etiology*:

SHEA'S CLASSIFICATION SYSTEM OF ULNAR INJURIES AT GUYON'S CANAL	
Type I	Involvement of the deep ulnar branch, hypothenar, and sensory
Type II	Involvement of deep ulnar motor branches
Type III	Involvement of the superficial ulnar sensory branch

Note: There can also be involvement of the ulnar nerve proximal to or at Guyon's canal to involve all three branches: the hypothenar, deep ulnar motor, and superficial ulnar sensory branches.

  - The nerve can be injured by cycling activities (cyclist's palsy), wrist ganglions, or rheumatoid arthritis (RA), trauma, or carpal bone fracture.
- *Clinical presentation*:
  - The muscles involved include all the ulnar nerve innervated intrinsic muscles of the hand (see box on top of the next page).
  - The patient may complain of painless wasting of the first DI. A severe claw hand may occur (lumbrical weakness) while the FDP remains intact, causing marked finger flexion.

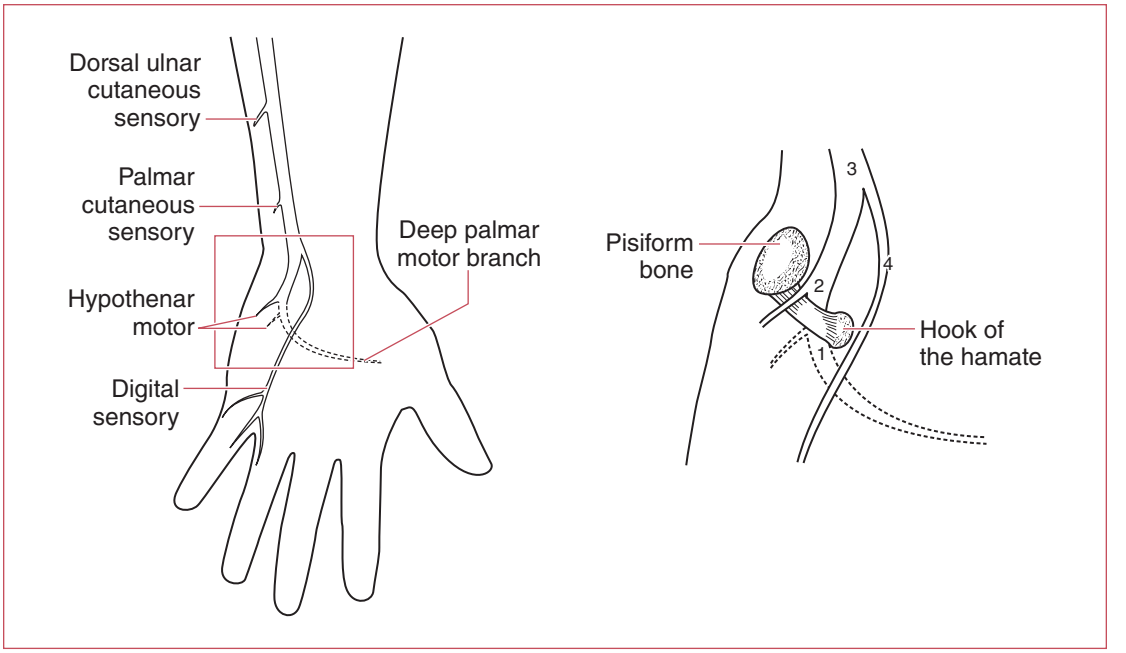


FIGURE 5–96 Detailed anatomy of the ulnar nerve at the wrist. Entrapment of the ulnar nerve at the wrist can take on several patterns: (1) pure motor affecting only the deep palmar motor branch, (2) pure motor affecting the deep palmar and hypothenar motor branches, (3) motor and sensory (proximal canal lesion), and rarely (4) pure sensory involving only the sensory fibers to the volar fourth and fifth fingers.

Muscles Involved		
4 Dorsal interossei 3 Palmar interossei 2 Lumbricals 1 Adductor pollicis 1/2 Flexor pollicis brevis	and the	Hypothenar eminence: • Opponens digiti minimi • Abductor digiti minimi • Flexor digiti minimi

Note: Compression of the ulnar nerve at Guyon's canal may spare the hypothenar muscle.

- *EDX findings*:
  - **NCS:**
    - SNAP: Ulnar SNAP to the fifth digit is abnormal, but DUC nerve SNAP is spared, as the DUC nerve is proximal to lesion.
    - CMAP: Abnormal (depends on which branch[es] are affected)
  - **EMG:** Abnormal activity in the ulnar nerve innervated hand muscles
- *Treatment*:
  - Interventions may include rehabilitation or a surgical procedure.

RADIAL NERVE

Anatomy

- **Origin (Figure 5–97):**
  - These nerve fibers originate from the C5–T1 roots. They continue on to contribute to the upper, middle, and lower trunks, posterior cord, and finally form the radial nerve.
- **Course:**

ARM

- The nerve is located posterior to the axillary artery. It descends between the long and medial heads of the triceps muscle toward the spiral groove.
  - Structures innervated *above* the spiral groove:
    - Triceps brachii
    - Anconeus
    - Posterior cutaneous nerve
    - Lower lateral cutaneous nerve
  - 🕮 Structures innervated *below* the spiral groove:
    - BR
    - Extensor carpi radialis-longus (ECR-L)
    - Posterior cutaneous nerve of forearm
- 10 cm proximal to the lateral epicondyle of the humerus, the nerve pierces the lateral intermuscular septum and enters the anterior compartment of the arm. It continues distally between the brachioradialis and brachialis.
- 🕮 At the lateral epicondyle, it then splits into a motor (posterior interosseus nerve [PIN]) and sensory branch (superficial radial nerve)

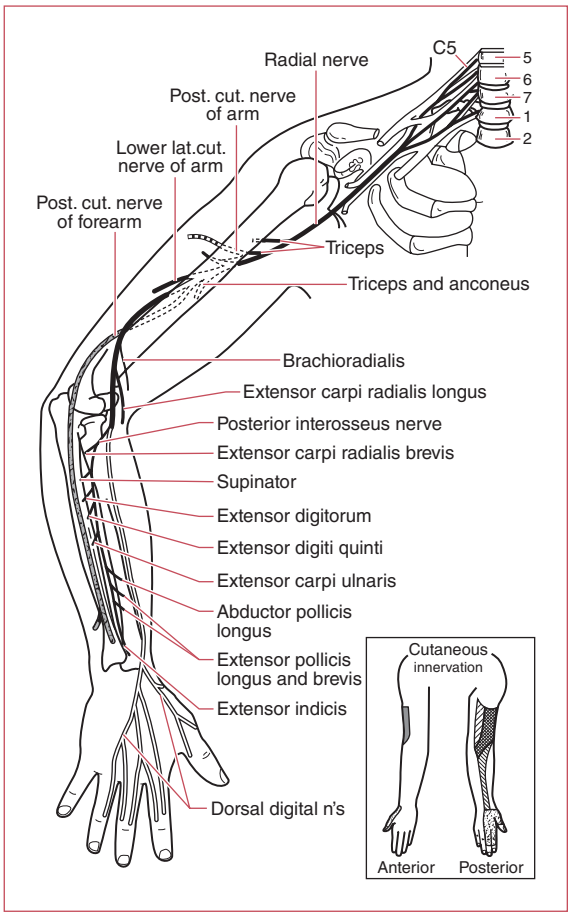


FIGURE 5–97 The radial nerve.

ELBOW

- **Superficial radial nerve**
- **PIN** innervates:
  - Extensor carpi radialis brevis (ECR-B)
  - Supinator
  - EDC
  - Extensor digiti minimi (EDM)
  - Extensor carpi ulnaris (ECU)
  - Abductor pollicis longus (APL)
  - Extensor pollicis longus (EPL)
  - Extensor pollicis brevis (EPB)
  - Extensor indicis proprius (EIP)

Injuries

AXILLA

CRUTCH PALSY

- *General*:
  - Nerve injury at this level can involve the posterior cord of the brachial plexus, most commonly affecting the radial nerve.
- *Etiology*:
  - The radial nerve or posterior cords of the brachial plexus can become compressed with improper axillary crutch use.
- *Clinical presentation*:
  - The patient may complain of weakness in all radial nerve innervated muscles, including the triceps brachii. Sensation may be decreased over the posterior arm and forearm.
- *EDX findings*:
  - **NCS:**
    - Abnormal radial SNAP and CMAPs
    - Injury to the posterior cords of the brachial plexus, which can affect the radial, axillary, and/or suprascapular nerves, can result in abnormal SNAPs and CMAPs of the affected nerves.
  - **EMG:**
    - Abnormal activity in all radial nerve innervated muscles
    - Injury to the posterior cords will result in abnormal activity in corresponding muscles that are innervated by the injured posterior cords.
- *Treatment*:
  - Interventions may focus on rehabilitation, discontinuing crutch use, static cock-up splint, or dynamic splinting

ARM

SPIRAL GROOVE

- *General*:
  - Injury at this site is also known as Saturday night palsy or honeymooner's palsy.
- *Etiology*:
  - The most common cause of radial nerve injury at the spiral groove is trauma, notably with humeral fractures.
  - Other common mechanisms of radial nerve injury at the spiral groove include iatrogenic injury (upper limb surgery, blood pressure cuff used during surgery), compression ("Saturday night palsy"—Figure 5–98), and intramuscular (IM) injection.

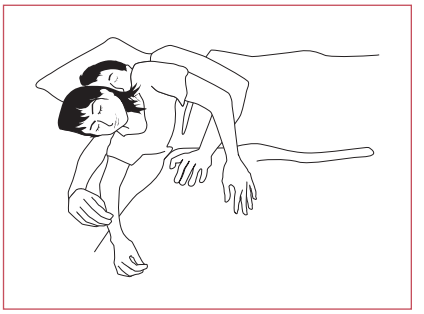


FIGURE 5–98 Mechanism of honeymooner's palsy (also known as Saturday night palsy).

- *Clinical presentation*:
  - Weakness noted in radial nerve innervated muscles below the spiral groove. Triceps brachii and anconeus are spared due to innervation above the spiral groove.
  - The patient complains of weakness of elbow flexion (BR weakness), supination (supinator weakness), wrist drop (ECR-L, ECR-B, and ECU weakness), and finger extension (EDC weakness). There is preservation of elbow extension (triceps, anconeus). Sensory deficits may occur in the dorsal aspect of the hand and posterior forearm.
- *EDX findings*:
  - **NCS:** Abnormal radial SNAPs and CMAP
  - **EMG:** Abnormal activity in all radial nerve innervated muscles below the spiral groove. The triceps is innervated proximal to the spiral groove, so the triceps should be normal.
- *Treatment*:
  - Intervention is primarily focused on rehabilitation. Surgery is indicated in cases of nerve transection, fractures, or open injuries.

Differential Diagnosis of a Wrist Drop
Mononeuropathy: PIN, radial nerve
Radiculopathy: C6 or C7
Diffuse polyneuropathy: Lead
Plexopathy: Posterior cord, upper trunk, and middle trunk
Central: SCI/TBI/CVA, and so forth

CVA, cerebrovascular accident; PIN, posterior interosseus nerve; SCI, spinal cord injury; TBI, traumatic brain injury.

FOREARM

RADIAL TUNNEL SYNDROME

- *General*:
  - The anatomic course of the radial nerve at the elbow is highly variable. It typically runs through an intramuscular septum between the brachialis and BR. Branching of the radial nerve into the PIN and superficial radial branches can occur before or after it travels through the intramuscular septum.
- *Etiology*:
  - The radial nerve can be entrapped between the brachialis and BR in the radial tunnel in the elbow.
- *Clinical presentation*:
  - Patients typically present with lateral proximal forearm pain that worsens with activity, which can mimic lateral epicondylitis.
  - Patients with radial tunnel syndrome will present with pain in the area radial tunnel, which is approximately 3 to 4 cm distal to the lateral epicondyle. In contrast, pain in lateral epicondylitis will be localized to the lateral epicondyle, where the ECR-B inserts (Bevelaqua et al., 2012).

RADIAL TUNNEL SYNDROME	LATERAL EPICONDYLITIS (TENNIS ELBOW)
Reproduction of symptoms with: • Resisted extension of the third digit during elbow extension • Resisted supination • Palpation of the radial tunnel	Reproduction of symptoms with: • Palpation directly on the lateral epicondyle • Cozen's test: Pain on resisted wrist extension • Less pain on resisted supination

- *EDX findings*:
  - NCS and needle EMG studies are typically normal.
- *Treatment*:
  - Interventions may include rehabilitation or surgical decompression.

POSTERIOR INTEROSSEUS NERVE SYNDROME (SUPINATOR SYNDROME, ARCADE OF FROHSE SYNDROME)

- *General*:
  - This is a lesion of the PIN, which is a motor nerve branch from the radial nerve.
  - It is considered a purely motor syndrome.
- *Etiology*:
  - The nerve can be injured by compression of the nerve at the *Arcade of Frohse* of the supinator (**Figure 5–99**). It can also be injured by a lipoma, ganglion cyst, synovitis from RA, or a 🕮 *Monteggia fracture*. This is a fracture of the proximal one-third of the ulna and dislocation of the radial head. It typically occurs from a fall on an outstretched hand with the forearm locked in full pronation.
- *Clinical presentation*:
  - PIN syndrome presents with painless weakness of all the PIN innervated distal extensors: EDC, EIP, ECU, EPB, EPL, and APL.
  - The PIN syndrome may spare the supinator, but always spares the BR, triceps, ECR-L, ECR-B, and anconeus, which are innervated proximally to the region of PIN entrapment.
  - A pseudo claw-hand deformity may be demonstrated (finger extensor weakness). Radial deviation is noted with wrist extension (ECU weakness) and sensation is spared.
- *EDX findings*:
  - **NCS:** Normal radial nerve SNAP. Abnormal radial CMAP when recording over PIN innervated muscles.
  - **EMG:** Abnormal activity in the muscles innervated by the PIN, which may include the supinator. Radial nerve innervated muscles proximal to the PIN lesion are spared.
- *Treatment*:
  - Interventions may include rehabilitation or surgical release.

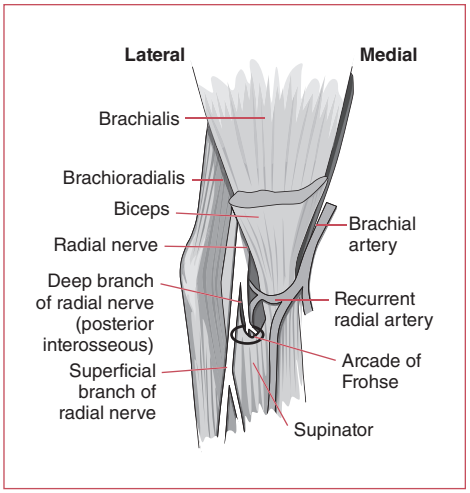


FIGURE 5–99 The Arcade of Frohse.

SUPERFICIAL RADIAL NEUROPATHY (WARTENBERG SYNDROME; FIGURE 5–100)

- *General*: Also known as Wartenberg syndrome or cheiralgia paresthetica. It is also known more informally as wristwatch syndrome.
- *Etiology*: Injury/compression of the superficial radial nerve at the wrist from a tight wristwatch, tight handcuffs, peripheral IV placement, and so forth.
- *Clinical presentation*:
  - This is a pure sensory syndrome with no muscle involvement. The patient may complain solely of sensory abnormalities including numbness, burning, or tingling on the dorsal radial aspect of the hand. Discomfort may be exacerbated with palmar and ulnar wrist flexion or forced pronation.

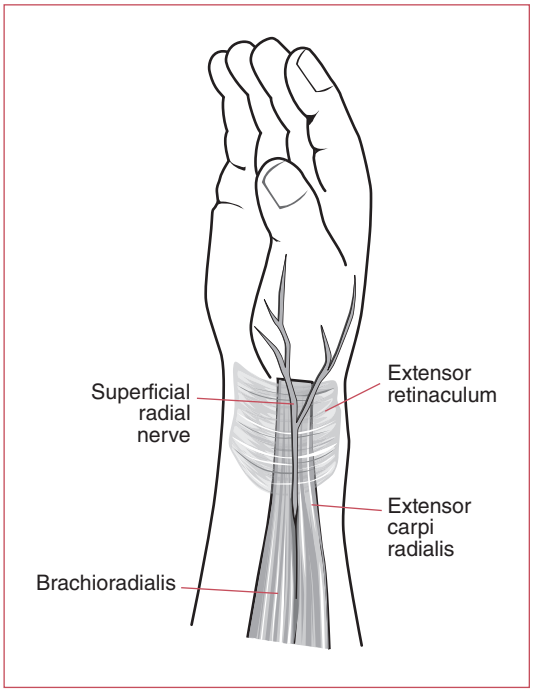


FIGURE 5–100 Superficial radial neuropathy injury.

- *EDX findings*:
  - **NCS:** Abnormal radial nerve SNAP but normal CMAP
  - **EMG:** Normal
- *Treatment*:
  - Interventions may include removal of the compressive irritant.

MUSCULOCUTANEOUS NERVE

Anatomy

- **Origin (Figure 5–101):**
  - These nerve fibers originate from the C5, C6, and C7 spinal nerve roots. These roots continue on to contribute to the upper trunk and lateral cord to finally form the musculocutaneous branch.
- **Course:**

ARM

- The nerve passes along the medial aspect of the humerus and innervates the:
  - Coracobrachialis
  - Biceps brachii
  - Brachialis (also innervated by the radial nerve)

FOREARM

- It continues anterior to the antecubital fossa (lateral to the biceps tendon) to provide cutaneous sensory innervation of the lateral forearm as the lateral antebrachial cutaneous nerve.

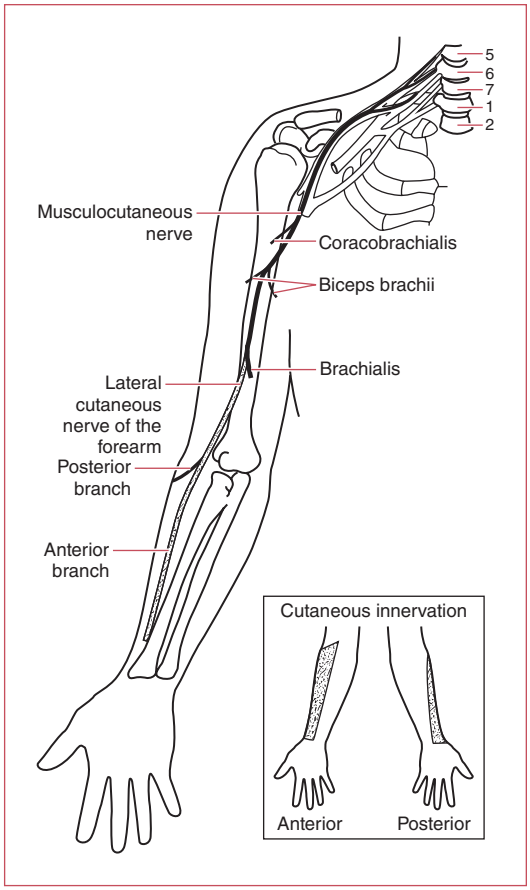


FIGURE 5–101 The musculocutaneous nerve.

Injury

MUSCULOCUTANEOUS NEUROPATHY

- *General*: Injury to the distal portion of the nerve is more common than to the proximal portion.
- *Etiology*:
  - The nerve can be injured by entrapment proximally from the coracobrachialis or distally, where it runs superficially. It can also be injured by trauma (proximal humeral fractures, shoulder dislocation, gunshot wounds), compression, and phlebotomy.
- *Clinical presentation*:
  - Involves musculocutaneous nerve innervated muscles including biceps and brachialis. The coracobrachialis is typically spared.
  - The patient may complain of elbow flexion weakness (biceps, brachialis weakness) and abnormal sensation over the lateral forearm.
- *EDX findings*:
  - **NCS:**
    - Abnormal SNAP in the lateral antebrachial cutaneous nerve
    - Abnormal CMAP to the biceps brachii
  - **EMG:**
    - Abnormal activity in the brachialis and biceps brachii
- *Treatment*:
  - Interventions may include rehabilitation or surgical release.

AXILLARY NERVE

Anatomy

- **Origin (Figure 5–102):**
  - These nerve fibers originate from the C5 and C6 spinal nerve roots. They continue on to contribute to the upper trunk and posterior cord to finally form the axillary nerve.
- **Course:**
  - The nerve passes inferior to the glenohumeral joint, through the quadrilateral space of the axilla and to the posterior aspect of the humerus. It innervates the:
    - Teres minor
    - Deltoid
    - Upper lateral cutaneous nerve

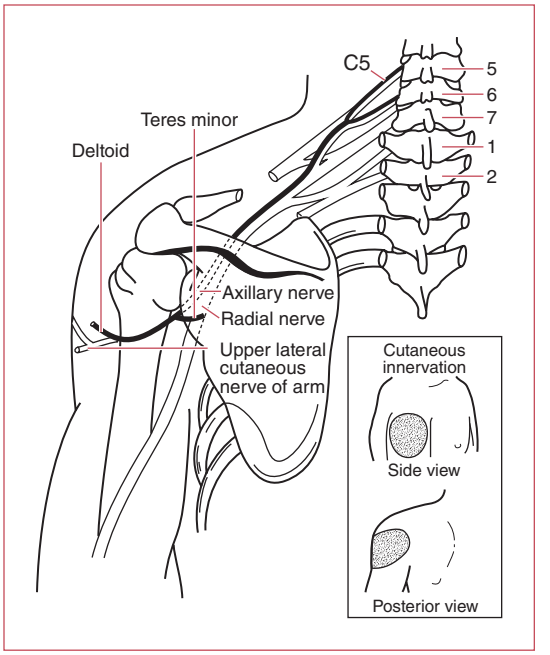


FIGURE 5–102 The axillary nerve.

Injury

AXILLARY NEUROPATHY

- *General*:
  - The nerve runs through the quadrangular space. This is bordered by the humerus, long head of the triceps brachii, teres minor, and teres major.
- *Etiology*:
  - The nerve can be injured by traction or compression from a shoulder dislocation, humeral head fracture, or improper axillary crutch use.
- *Clinical presentation*:
  - Involves all axillary nerve innervated muscles (deltoid and teres minor)
  - The patient may complain of weakness of shoulder flexion and abduction (deltoid weakness) and external rotation (teres minor weakness). There may also be abnormal sensation of the lateral shoulder.
- *EDX findings*:
  - **NCS:**
    - SNAP not available
    - CMAP: Abnormal
  - **EMG:**
    - Abnormal activity in the axillary nerve innervated muscles (deltoid, teres minor). The anterior and lateral heads of the deltoid show abnormalities more commonly than the posterior head.
- *Treatment*:
  - Interventions may include rehabilitation or surgical decompression, as well as discontinuation of crutch use.

SUPRASCAPULAR NERVE

Anatomy

- **Origin (Figure 5–103):**
  - These nerve fibers originate from the C5 and C6 spinal nerve roots. They continue on to contribute to the upper trunk, which branches off to form the suprascapular nerve.
- **Course:**

NECK

- It passes the posterior triangle of the neck and runs beneath the trapezius to the superior margin of the scapula.

- 🕮 It runs through the *suprascapular* notch, which is covered by the transverse scapular ligament and branches to innervate the supraspinatus. The nerve then wraps around the *spinoglenoid* notch to innervate the infraspinatus.

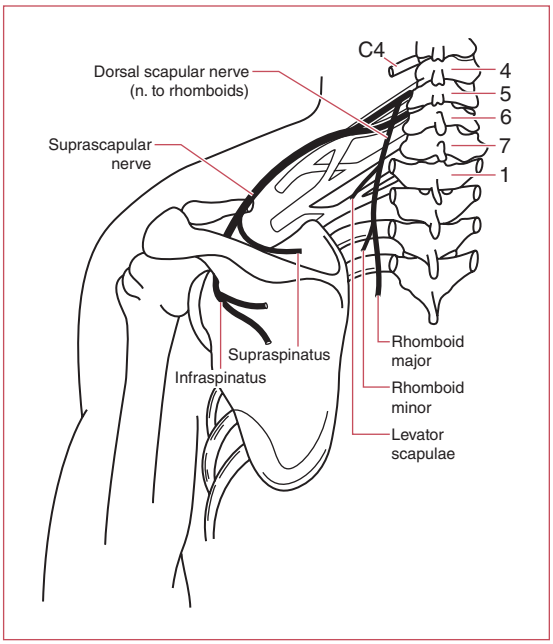


FIGURE 5–103 The suprascapular nerve.

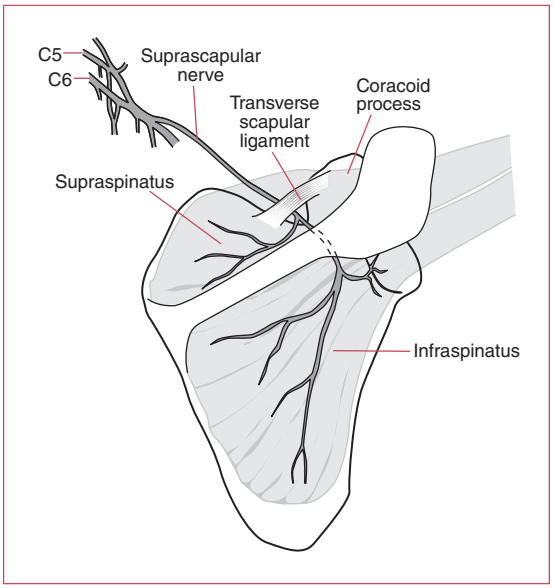


FIGURE 5–104 Suprascapular nerve injury sites.

Injury

🕮 ***SUPRASCAPULAR NEUROPATHY***

- *General*:
  - This is the only peripheral nerve injury at the trunk level. It is the most commonly involved nerve in neuralgic amyotrophy (**Figure 5–104**).
- *Etiology*:
  - The nerve can be injured from trauma, including forced scapular protraction, penetrating wounds, traction from a massive rotator cuff tear, stinger/Erb's palsy; compression from spinoglenoid ganglions, hematoma, suprascapular or spinoglenoid notch entrapment, or paralabral cyst.
  - Activities involving exaggerated shoulder movements, including sports with repetitive overhead throwing/hitting such as volleyball, baseball, and lacrosse, may also injure the nerve. Volleyball more commonly injures branches to the infraspinatus muscle.
- *Clinical presentation*:
  - Injury to the nerve at the suprascapular notch results in weakness in both supraspinatus and infraspinatus muscles. Nerve injury at the spinoglenoid notch will result in weakness only in the infraspinatus muscle.
  - The patient may present with weakness in abduction (supraspinatus) and/or external rotation (infraspinatus) of the glenohumeral joint.
- *EDX findings*:
  - **NCS:**
    - SNAP not available
    - CMAP: Abnormal
  - **EMG:**
    - Abnormal activity in the infraspinatus (IS) only if entrapment is at the spinoglenoid notch or both supraspinatus and infraspinatus muscles if nerve entrapment is at the suprascapular notch.
- *Treatment*:
  - Interventions may include rehabilitation, paralabral cyst aspiration, or surgical decompression.

LONG THORACIC NERVE

Anatomy

- **Origin (Figure 5–105):**
  - These nerve fibers originate directly from the C5, C6, and C7 spinal nerve roots.
- **Course:**

NECK

- The nerve runs distally along the thoracic wall to innervate the serratus anterior (SALT—Serratus Anterior Long Thoracic).

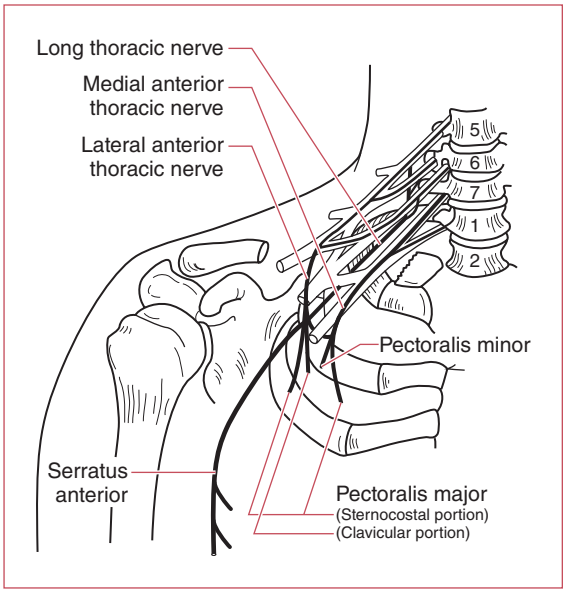


FIGURE 5–105 The long thoracic nerve.

Injury

SCAPULAR WINGING

- *General*:
  - The long thoracic nerve innervates the serratus anterior, which protracts and laterally rotates the scapula.
  - The spinal accessory nerve innervates the trapezius, which retracts, elevates, and medially rotates the scapula.
  - The serratus anterior and trapezius help to balance scapular motion in opposing directions. Dysfunction in one of the muscles due to nerve injury can result in scapular winging.
  - There are two main types of shoulder winging that should be differentiated from each other (Table 5–31):
    - **Medial winging** is due to serratus anterior weakness from a long thoracic nerve injury.
    - **Lateral winging** results from trapezius weakness from a spinal accessory nerve injury.
- *Etiology*:
  - The nerve can be injured by traction from a fall, MVA, sports activities, or shoulder bags.
- *Clinical presentation*:
  - Shoulder pain and weakness as presenting symptoms with scapular winging.

TABLE 5–31 Scapular Winging: Medial Versus Lateral Scapular Winging

	MEDIAL SCAPULAR WINGING	LATERAL SCAPULAR WINGING
Nerve injured	Long thoracic nerve	Spinal accessory nerve
Affected muscle/function	Serratus anterior (Normal function: protracts and laterally rotates the scapula)	Trapezius (Normal function: retracts and medially rotates the scapula)
Position of scapula (Medial border)	Scapula retracted and elevated Medial scapular border closer to midline due to unopposed retraction from the trapezius	Scapula protracted and depressed Medial scapular border is pulled away from midline by unopposed protraction from the serratus anterior
Shoulder abduction causes	Decreases winging	Increases winging

- *EDX findings*:
  - **NCS:**
    - SNAP: Not available
    - CMAP: Abnormal

- **EMG:**
  - Abnormal activity in the serratus anterior with a long thoracic nerve injury. Abnormal activity in the trapezius and sternocleidomastoid (SCM) indicates injury to the spinal accessory nerve.
- *Treatment*:
  - Interventions may include rehabilitation or surgical procedure.
  - **Rehabilitation of serratus anterior injury/injury to the long thoracic nerve:** (a) acute stage, pain reduction and ROM exercise; (b) intermediate stage, passive stretching of the rhomboids, levator scapulae, and pectoralis minor; (c) late stage, strengthening exercise of all shoulder girdle muscles, including the trapezius.
  - **Rehabilitation of trapezius palsy/injury to the spinal accessory nerve:** Will involve physical therapy to adequately strengthen adjacent muscle groups, including rhomboids and levator scapulae.
  - Surgical repair with a dynamic muscle transfer is recommended if the patient fails conservative treatment.

LOWER LIMB MONONEUROPATHY

LATERAL CUTANEOUS NERVE OF THE THIGH (LATERAL FEMORAL CUTANEOUS NERVE)

Anatomy

- **Origin (Figure 5–106):**
  - These pure sensory nerve fibers originate from the L2 and L3 nerve roots from the posterior divisions of the lumbar plexus.
- **Course:**

PELVIS/THIGH

- The nerve continues distally to pass over the iliacus toward the anterior iliac spine. It passes underneath the inguinal ligament to provide sensation to the lateral thigh.

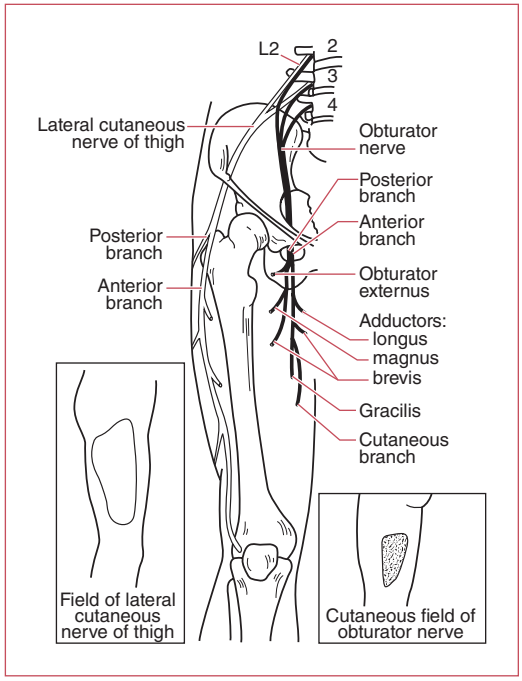


FIGURE 5–106 The lateral cutaneous nerve of the thigh (lateral femoral cutaneous nerve).

Injury

LATERAL CUTANEOUS NERVE OF THE THIGH NEUROPATHY (LATERAL FEMORAL CUTANEOUS NEUROPATHY; MERALGIA PARESTHETICA)

- *General*:
  - Lateral femoral cutaneous neuropathy is also known as meralgia paresthetica and is mainly a clinical diagnosis.
- *Etiology*:
  - The nerve can be injured from chronic compression by a repeated low-grade trauma, from a protuberant abdomen, pregnancy, or tight clothing.
  - Diabetes, tumor, infection, and rapid weight gain or weight loss can also affect the nerve.
  - Iatrogenic cause may be present, including incisions for lower abdominal/pelvic surgeries and laparoscopic hernia repairs (Ivins, 2000).
- *Clinical presentation*:
  - This is a pure sensory syndrome with no muscle involvement.

  - The patient may report sensory complaints in the lateral thigh, including pain, numbness, burning, or a dull ache, exacerbated with hip extension or hyperflexion, prolonged squatting, or driving.
- *EDX findings*:
  - **NCS:** Abnormal lateral cutaneous nerve of the thigh (lateral femoral cutaneous nerve) SNAP. CMAP not available.
  - **EMG:** Not available
- *Treatment*:
  - Interventions may include rehabilitation, NSAIDs, cortisone injections, and surgical release.
  - Symptoms may be self-limited.
  - In addition, removal of compressive clothing should occur (e.g., wide belt, compressive athletic clothing).

FEMORAL NERVE

Anatomy

- **Origin (Figure 5–107):**
  - These nerve fibers originate from the L2, L3, and L4 spinal nerve roots, continue on as the posterior division of the lumbar plexus, and terminate as the femoral nerve.
- **Course:**
  - The nerve runs through the psoas muscle. It travels under the inguinal ligament lateral to the femoral artery and through the femoral triangle to innervate the following muscles:
    - Iliacus
    - Pectineus (1/2)
    - Sartorius
    - Quadriceps muscles:
      - Rectus femoris
      - Vastus lateralis
      - Vastus intermedius
      - Vastus medialis
  - The **saphenous nerve**, a pure sensory nerve, branches from the femoral nerve and travels through the adductor canal to the rest of the leg and provides sensation to the anteromedial thigh, calf, and foot.

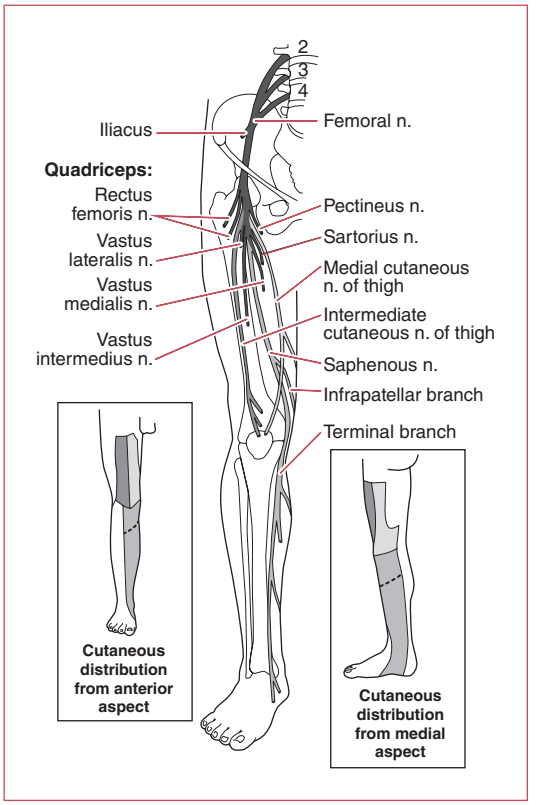


FIGURE 5–107 The femoral nerve.
n, nerve.

Injury

FEMORAL NEUROPATHY

- *General*:
  - This nerve is the largest branch of the lumbar plexus.
- *Etiology*:
  - The nerve can be injured by trauma (e.g., fracture, abdominal/pelvic surgery, anterior total hip arthroplasty, direct trauma from cardiac catheterization), compression from a retroperitoneal hematoma, tumor, or inguinal ligament.
  - The most common cause is iatrogenic from abdominal or pelvic surgery.
- *Clinical presentation*:
  - Involved muscles include all femoral nerve innervated muscles. The patient may complain of weakness of knee extension (quadriceps), knee instability, and decreased sensation over the anterior thigh and medial leg. Hip flexion weakness is noted with injuries occurring above the inguinal ligament.

- *EDX findings*:
  - **NCS:**
    - Abnormal saphenous nerve SNAP
    - Abnormal CMAP to the rectus femoris
  - **EMG:**
    - Abnormal activity in the femoral nerve innervated muscles. Involvement of obturator nerve innervated muscles (e.g., adductor longus) and/or lumbar paraspinals could indicate an L3/L4 radiculopathy over a femoral neuropathy.
- *Treatment*:
  - Interventions may include rehabilitation or surgical procedure.

DIABETIC AMYOTROPHY

- 🕮 *General*:
  - Diabetic amyotrophy is the most common cause of femoral neuropathy.
  - Diabetic amyotrophy is an episodic, monophasic, asymmetrical, proximal diabetic neuropathy distinct from other types of distal diabetic peripheral neuropathies.
  - It is also known as lumbosacral radiculoplexus neuropathy, as it can involve the plexus and nerve roots, as well as peripheral nerves. It predominantly affects the lumbosacral plexus.
- *Etiology*:
  - There is increasing evidence for an immune-mediated inflammatory microvasculitis, with clear association to diabetes, but an unclear link in its pathophysiology.
  - 🕮 Nerve biopsy shows degenerative changes secondary to microvasculitis with accompanying ischemia and inflammation, further ratified by cerebrospinal fluid (CSF) findings demonstrating elevated protein.
- Clinical presentation (Pasnoor et al., 2013):
  - It typically affects an older group of patients with diabetes, more frequently males, usually over age 50, with a low body mass index (BMI). Most patients have type 2 diabetes mellitus.
  - This neuropathy begins with severe unilateral pain in the lumbar region or proximal lower extremity, which commonly spreads to the contralateral side within weeks to months. Patients then develop weakness and atrophy of the proximal > distal lower extremity musculature.
  - There is typically associated weight loss that precedes symptoms.
  - The patient may complain of asymmetric thigh pain, knee extension weakness (quadriceps), and atrophy. Loss of the patellar reflex may also occur.
- *EDX findings*:
  - **NCS:** Asymmetrically reduced sensory and motor AP amplitude in affected muscles
  - **EMG:** Abnormal activity seen in femoral innervated muscles (±), adductors, iliopsoas, and paraspinals
- *Treatment*:
  - Diabetic amyotrophy is a self-limited condition, but the recovery process is gradual and occurs over a period of months. There are some patients who are left with residual lower extremity weakness.
  - Treatment is focused around pain control and optimizing glycemic control. Physical therapy can assist in improving strength and functional mobility.

SAPHENOUS NEUROPATHY

- *General*:
  - This nerve is the largest and longest sensory branch of the femoral nerve.
  - It supplies sensation to the medial aspect of the leg, the medial malleolus, and medial arch of the foot.
- *Etiology*:
  - The nerve can be entrapped in the hip adductor canal (aka subsartorial or Hunter's canal) or between the sartorius and gracilis.
  - It can also be related to iatrogenic trauma, including knee arthroscopy, and meniscectomy or vascular surgery, such as catheterization or thrombectomy.
- *Clinical presentation*:
  - This is a pure sensory syndrome with no muscle involvement.

  - The patient may complain of medial knee pain (infrapatellar branch) with abnormal sensation radiating distally along the medial aspect of the leg and foot.
- *EDX findings*:
  - **NCS:** Abnormal saphenous nerve SNAP. Normal femoral nerve CMAP.
  - **EMG:** Normal EMG of femoral nerve innervated muscles.
- *Treatment*:
  - Interventions may include rehabilitation or surgical procedure.

OBTURATOR NERVE

Anatomy

- **Origin (Figure 5–108):**
  - Nerve fibers originating from the L2, L3, and L4 spinal nerve roots continue on as the anterior portion of the lumbar plexus in front of the sacroiliac joint to form the obturator nerve, which innervates the hip adductor musculature.
- **Course:**
  - The nerve passes through the psoas major and obturator foramen to innervate:
    - Pectineus (1/2)
    - Adductor brevis
    - Adductor longus
    - Adductor magnus
    - Obturator externus
    - Gracilis
    - Cutaneous branch

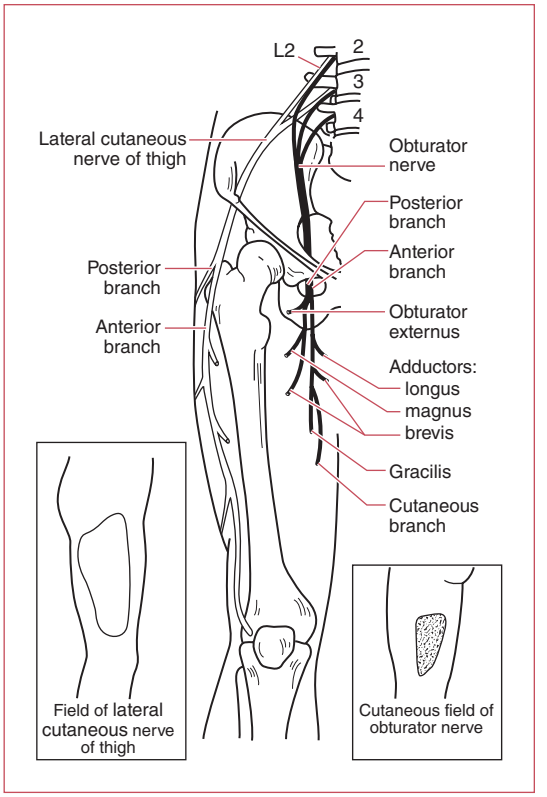


FIGURE 5–108 The obturator nerve.

Injury

OBTURATOR NEUROPATHY

- *General*:
  - This nerve injury can occur in conjunction with a femoral nerve injury.
  - The obturator nerve gives sensation to the medial aspect of the thigh and innervates the muscles in the medial compartment of the thigh, which includes the obturator externus, adductor brevis, adductor longus, adductor magnus muscles, and gracilis muscles.
- *Etiology*:
  - The nerve can be injured by surgery, compression from a hematoma or tumor, a pelvic fracture, or obturator hernia.
- *Clinical presentation*:
  - Can involve all muscles innervated by the obturator nerve.
  - The most common symptom is altered sensation in the inguinal/medial thigh region, including pain, numbness, and/or paresthesias.
  - Patients are also usually noted to have weakness with hip ADduction and internal rotation. During ambulation, they present with the hip externally rotated and abnormally abducted, which results in a wide-based, circumducting gait.
- *EDX findings*:
  - **NCS:** None available
  - **EMG:** Abnormal activity in the obturator nerve innervated muscles
- *Treatment*:
  - Intervention may include rehabilitation or surgical repair.

SCIATIC NERVE

Anatomy

- **Origin (Figure 5–109):**
  - These nerve fibers from the L4, L5, S1, S2, and S3 roots travel through the anterior and posterior divisions of the sacral plexus and eventually form the sciatic nerve, which travels posteriorly in the leg.
- **Course:**
  - 🕮 The sciatic nerve exits the pelvis through the greater sciatic foramen between the lesser trochanter and ischial tuberosity. The sciatic nerve proper comprises a tibial (medial portion of the nerve) and peroneal (fibular; lateral portion of the nerve) division. It travels as one unit distally to the popliteal fossa, where it splits into its respective divisions: the peroneal (fibular) division and the tibial division.
  - 🕮 Sciatic nerve muscle innervation in the thigh.
  - The peroneal (fibular) division of the sciatic nerve innervates the short head of the biceps femoris.
  - The tibial division of the sciatic nerve innervates:
    - Long head of the biceps femoris
    - Semitendinosus
    - Semimembranosus
    - Adductor magnus (also innervated by the obturator nerve)

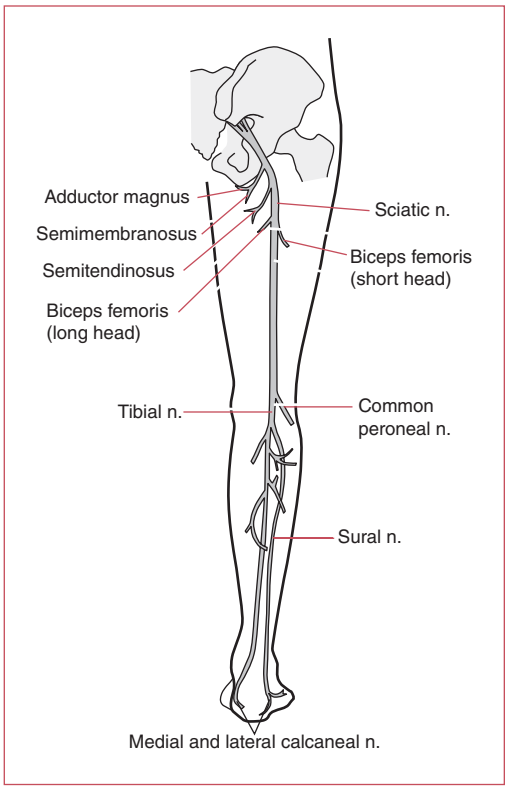


FIGURE 5–109 The sciatic nerve.
Note: The terms peroneal and fibular are used interchangeably. The word "peroneal" has been kept in the figure; however, please note that peroneal = fibular.
n, nerve.

Injury

SCIATIC NEUROPATHY

- *General*:
  - This is the largest nerve in the human body. The peroneal (fibular) portion makes up the outer two-thirds of the nerve proper.
- *Etiology*:
  - The nerve can be injured by hip trauma, hip replacement, injection, hematoma, pelvic fracture, penetrating wounds, or a gravid uterus.
  - 🕮 *Piriformis syndrome* is an uncommon type of compressive sciatic neuropathy at the pelvic outlet that mainly affects the peroneal (fibular) portion of the nerve as it runs inferior or through this muscle (**Figure 5–110**). While there is no consensus on diagnosing piriformis syndrome, its estimated incidence is approximately 4% (Hicks & Varacallo, 2019).

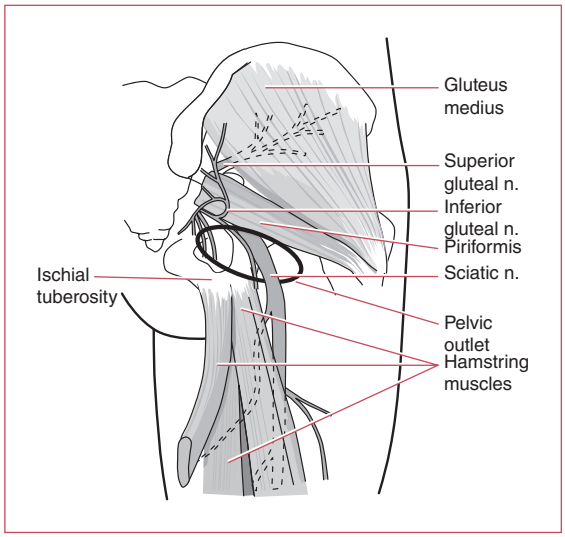


FIGURE 5–110 Piriformis syndrome.
n, nerve.

- *Clinical presentation*:
  - Can involve all muscles innervated by the sciatic nerve.
  - The patient's complaints depend on which portion of the nerve is more involved. It can present as weakness of knee flexion (hamstring weakness) and include muscles and cutaneous innervation of both the peroneal (fibular) and tibial nerves.
  - The peroneal (fibular) portion is more vulnerable to injury than the tibial because the peroneal (fibular) portion is more fixated in the pelvis. It has larger fascicles with less protective epineural tissue.
  - The lateral hamstring and Achilles reflexes may be abnormal.
- *EDX findings*:
  - **NCS:**
    - SNAP: Abnormal superficial peroneal (fibular) and sural sensory nerves SNAPs
    - Abnormal tibial and peroneal (fibular) nerve CMAPs
  - **EMG:**
    - Abnormal activity in all sciatic innervated musculature, including the short and long heads of the biceps femoris.
- *Treatment*:
  - Interventions may include rehabilitation or surgical decompression.

TIBIAL NERVE

Anatomy

- **Origin (Figure 5–111):**
  - These nerve fibers originate from the L4, L5, S1, and S2 (S3) spinal nerve roots. They continue on as part of the sciatic nerve and then branch at the popliteal fossa to finally form the tibial nerve.

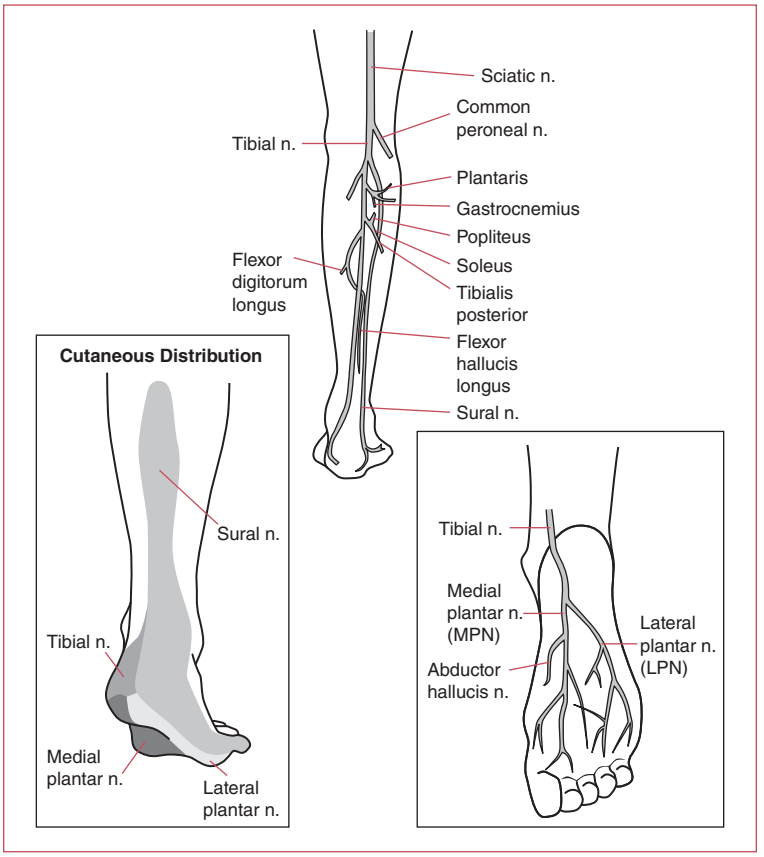


FIGURE 5–111 The tibial nerve. *Note*: The terms peroneal and fibular are used interchangeably. The word "peroneal" has been kept in the figure; however, please note that peroneal = fibular.
n, nerve.

- **Course:**
  - At the distal one-third of the thigh, the tibial portion of the nerve runs posterior to the knee and continues distally to innervate:
    - Plantaris
    - Medial and lateral gastrocnemius
    - Popliteus
    - Soleus
  - At the soleus, it continues on as the posterior tibial nerve and innervates:
    - Tibialis posterior (TP)
    - Flexor digitorum longus (FDL)
    - Flexor hallucis longus (FHL)
  - The nerve runs under the flexor retinaculum and divides into three branches.
    - *Medial plantar nerve*:
      - Abductor hallucis
      - Flexor digitorum brevis
      - Flexor hallucis brevis
      - Lumbrical
      - Sensory branch
    - *Lateral plantar nerve*:
      - Lumbricals
      - ADM
      - Quadratus plantae
      - Interossei
      - Adductor hallucis
      - Sensory branches
    - *Calcaneal nerve*:
      - Sensory branch

Injury

TARSAL TUNNEL SYNDROME

- *General*:
  - The tibial nerve may become entrapped in the tarsal tunnel (**Figure 5–112** and **Table 5–32**). This condition is relatively rare.
- *Etiology*:
  - The tibial nerve can be injured by compression under the flexor retinaculum in the medial ankle.
- *Clinical presentation*:
  - Involves all muscles innervated by the tibial nerve distal to the tarsal tunnel. The patient may complain of symptoms related to intrinsic foot weakness.
  - Perimalleolar pain, numbness, and paresthesias may extend to the toes and soles. Heel sensation may be spared due to the calcaneal branch departing proximal to the tunnel.
  - It is reproduced by ankle inversion. A positive Tinel's sign at the medial ankle may be elicited.
- *EDX findings*:
  - **NCS:**
    - SNAP: Abnormal plantar nerve. Normal calcaneal nerve.
    - CMAP: Abnormal medial and lateral plantar nerve

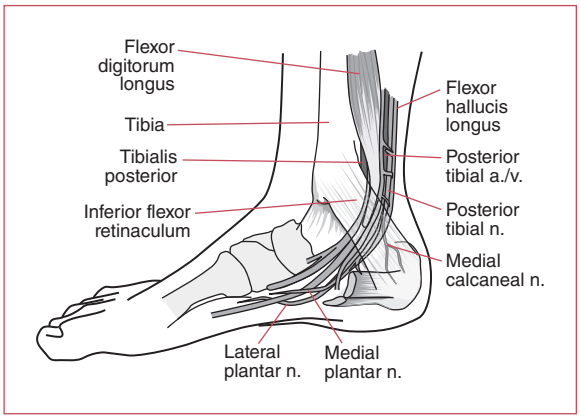


FIGURE 5–112 The posterior tarsal tunnel. a./v., artery/vein; n, nerve.

TABLE 5–32 Tarsal Tunnel Contents

Tom	**T**ibialis posterior
Dick	**F**lexor **d**igitorum longus
And	**P**osterior tibial **a**rtery
Very	**P**osterior tibial **v**ein
Nervous	**T**ibial **n**erve
Harry	**F**lexor **h**allucis longus

Note: Mnemonic for the contents of the tarsal tunnel is as follows: **T**om, **D**ick, **A**nd **V**ery **N**ervous **H**arry.

  - **EMG:**
    - Abnormal activity in the tibial nerve innervated muscles
- *Treatment*:
  - Interventions may include rehabilitation or surgical release.

COMMON PERONEAL (FIBULAR) NERVE

Anatomy

- **Origin:**
  - These nerve fibers originate from the L4–S2 spinal nerve roots. They continue on as the sciatic nerve, which divides to form the peroneal (fibular) nerve.
- **Course (Figures 5–113 and 5–114):**
  - In the posterior thigh, the peroneal (fibular) fibers within the sciatic nerve innervate the short head of the biceps femoris, the only peroneal (fibular) innervated muscle above the level of the fibular neck. More distally, the sciatic nerve bifurcates above the popliteal fossa into the common peroneal (fibular) and tibial nerve.

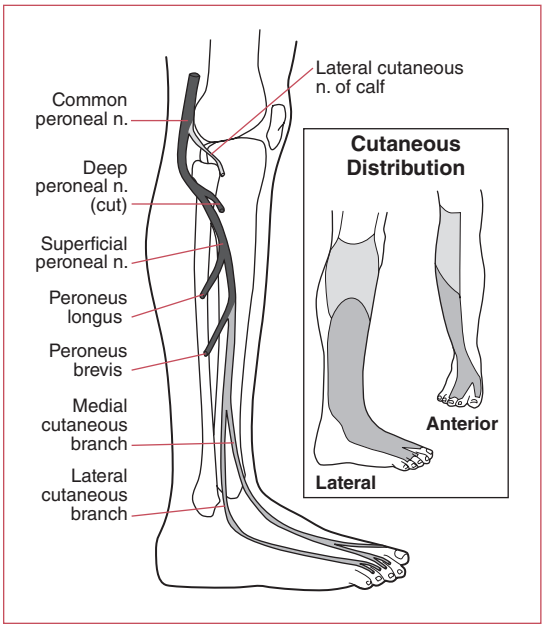


FIGURE 5–113 The superficial peroneal (fibular) nerve. *Note*: The terms peroneus and fibularis are used interchangeably. The words "peroneus" and "peroneal" have been kept in the figure; however, please note that peroneus = fibularis; peroneal = fibular.
n, nerve.

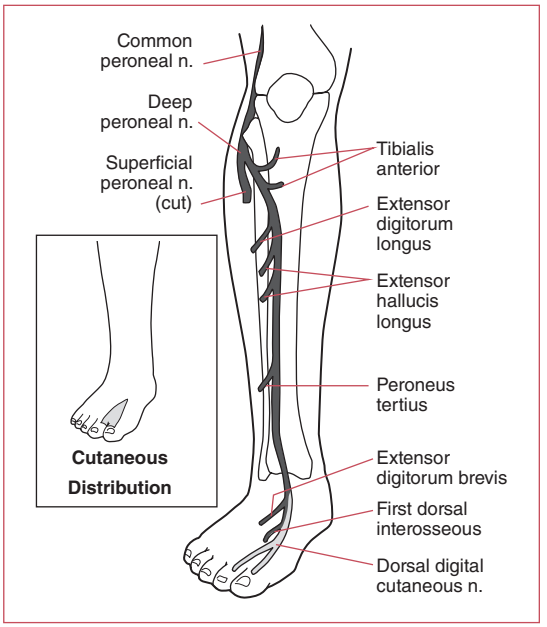


FIGURE 5–114 The deep peroneal (fibular) nerve. *Note*: The terms peroneus and fibularis are used interchangeably. The words "peroneus" and "peroneal" have been kept in the figure; however, please note that peroneus = fibularis; peroneal = fibular.
n, nerve.

Superficial Peroneal (Fibular) Nerve **innervates:**	Deep Peroneal (Fibular) Nerve **innervates:**
• Peroneus (fibularis) longus • Peroneus (fibularis) brevis • Medial cutaneous nerve • Lateral cutaneous nerve	• Tibialis anterior • Extensor digitorum longus • Extensor hallucis longus • Peroneus (fibularis) tertius • Extensor digitorum brevis • First dorsal interossei • Dorsal distal cutaneous nerve

ACCESSORY PERONEAL (FIBULAR) NERVE

- *General* (**Figure 5–115**):
  - This is an anomalous nerve branch from the superficial peroneal (fibular) nerve that travels posterior to the lateral malleolus to innervate some or all of the extensor digitorum brevis (EDB).
  - Present in one-third of the population.
- *EDX findings*:
  - Lower peroneal (fibular) CMAP (recording at EDB) with stimulation at the ankle compared with stimulation at the knee, with greater amplitudes above the fibular head. The presence can be confirmed by stimulating behind the lateral malleolus, which will also elicit a CMAP from the EDB. With this anomalous innervation, the EDB can be spared even with a deep peroneal (fibular) nerve injury.

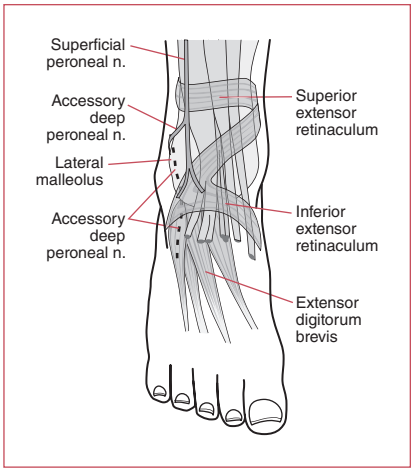


FIGURE 5–115 The accessory peroneal (fibular) nerve.
Note: The terms peroneal and fibular are used interchangeably. The word "peroneal" has been kept in the figure; however, please note that peroneal = fibular.
n, nerve.

Injury

📖 *COMMON PERONEAL (FIBULAR) NEUROPATHY*

- *General* (**Figure 5–116**):
  - The most common site of this nerve injury is at the fibular head.
- *Etiology*:
  - The nerve can be injured by compression from prolonged leg crossing, weight loss, poor positioning during surgery, poor cast application, prolonged squatting position (strawberry pickers' palsy), and metabolic disorders such as diabetes.
- 📖 *Clinical presentation*:
  - The muscles involved include all muscles supplied by the deep and superficial branches of the common peroneal (fibular) nerve (short head of the biceps femoris is spared).
  - The patient may complain of weakness of the dorsiflexors (tibialis anterior [TA], extensor digitorum longus [EDL], extensor hallucis longus [EHL]), resulting in a *foot drop* or foot slap and a *steppage gait*. Weakness of only the ankle dorsiflexors and ankle evertors helps to clinically differentiate a peroneal (fibular) nerve injury from an L5 radiculopathy. A radiculopathy will also involve the ankle invertors.
  - Sensory loss may be noted over the distribution of the deep and superficial peroneal (fibular) nerves.
  - A positive Tinel's sign may be noted at the fibular head.

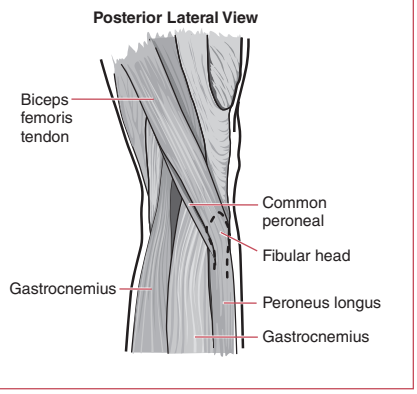


FIGURE 5–116 Common peroneal (fibular) nerve proximity to the fibula head.
Note: The terms peroneus and fibularis are used interchangeably. The words "peroneus" and "peroneal" have been kept in the figure; however, please note that peroneus = fibularis; peroneal = fibular.

- *EDX findings*:
  - **NCS:**
    - Abnormal superficial peroneal (fibular) SNAP
    - CMAP: Abnormal waveform change from below to above the fibular head. The TA can be used if the EDB is atrophied.
  - **EMG:**
    - Abnormal activity in the muscles innervated by the superficial and deep peroneal (fibular) nerves. The short head of the biceps femoris is spared (sciatic nerve, peroneal [fibular] division).
- *Treatment*:
  - Interventions may include rehabilitation or surgical procedure.

DEEP PERONEAL (FIBULAR) NEUROPATHY

- *General*:
  - Injury to this nerve is associated with anterior *tarsal tunnel syndrome* (**Figure 5–117**), which refers to entrapment of the deep peroneal (fibular) nerve under the inferior extensor retinaculum of the ankle.
- *Etiology*:
  - The terminal portion of the deep peroneal (fibular) nerve can be injured by compression from footwear (high boots, tight shoes, high-heeled shoes) or intrinsic etiologies (osteophyte, ganglion cyst, lipoma) as it passes under the extensor retinaculum, as well as trauma (ankle sprains or fractures).
- *Clinical presentation*:
  - The patient may complain of foot pain, weakness, and atrophy (EDB). There may also be numbness and paresthesias in the first and second web space. Pain may be located over the dorsum of the foot and relieved with motion.
- *EDX findings*:
  - **NCS:**
    - SNAP: Abnormal deep peroneal (fibular) SNAP (abnormal findings to the first web space)
    - CMAP: Abnormal peroneal (fibular) CMAP with recording at the EDB (abnormal findings to the EDB).
  - **EMG:** Abnormal activity in deep peroneal (fibular) innervated muscles
- *Treatment*:
  - Interventions may include rehabilitation or surgical resection.

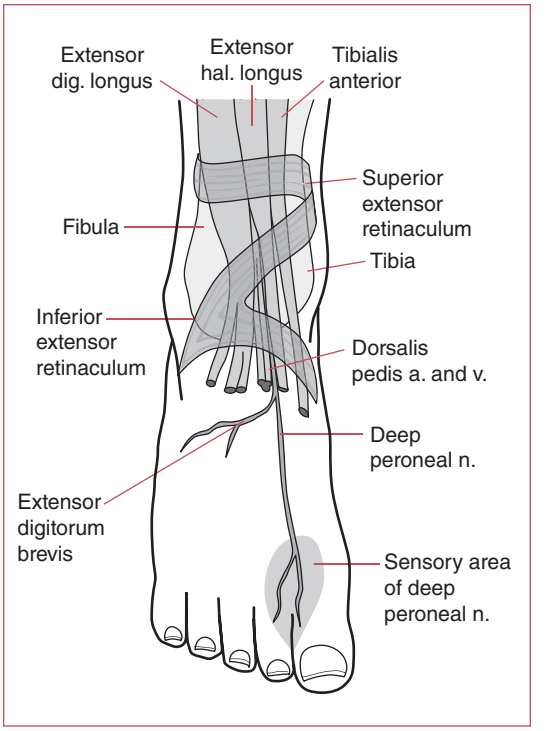


FIGURE 5–117 The anterior tarsal tunnel. *Note*: The terms peroneal and fibular are used interchangeably. The word "peroneal" has been kept in the figure; however, please note that peroneal = fibular. a, artery; n, nerve; v, vein.

SUPERFICIAL PERONEAL (FIBULAR) NEUROPATHY

- *General*:
  - After innervating the peroneus (fibularis) longus and brevis, the nerve continues distally as a pure sensory nerve.
- *Etiology*:
  - The nerve can be injured by compression from trauma, ankle sprain, compartment syndrome, or a lipoma.
- *Clinical presentation*:
  - Patients complain of pain, numbness, and/or paresthesias in the loss of sensation in distal anterolateral calf and dorsal foot (except for the first webspace). Foot eversion weakness can

be noted if the injury is proximal to the nerve innervating the peroneus (fibularis) longus and peroneus (fibularis) brevis muscles.

- *EDX findings*:
  - **NCS:**
    - Abnormal superficial peroneal (fibular) SNAP. Normal deep peroneal (fibular) SNAP.
    - Superficial peroneal (fibular) CMAP not available. Normal deep peroneal (fibular) CMAP.
  - **EMG:** Abnormal activity in the peroneus (fibularis) longus and/or peroneus (fibularis) brevis but normal findings in deep peroneal (fibular) nerve innervated muscles.
- *Treatment*:
  - Interventions may include rehabilitation or surgical release.

SURAL NERVE

Anatomy

- **Origin (Figure 5–118):**
  - These nerve fibers originate from branches of the tibial and common peroneal (fibular) nerves.
- **Course:**
  - The sural nerve travels from the proximal posterior calf to the lateral malleolus of the proximal ankle. It supplies cutaneous innervation to the lateral calf and foot.

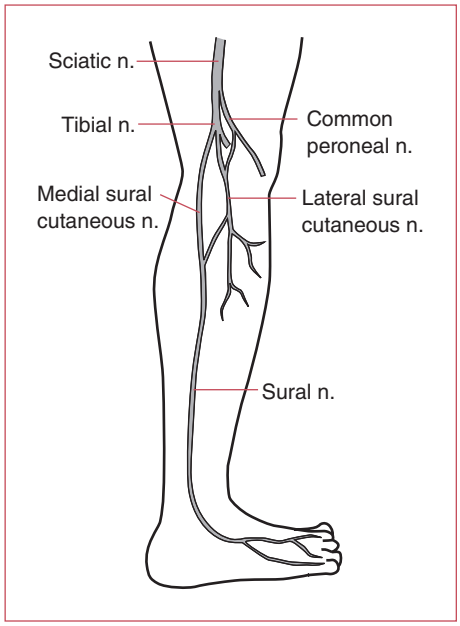


FIGURE 5–118 The sural nerve.
Note: The terms peroneal and fibular are used interchangeably. The word "peroneal" has been kept in the figure; however, please note that peroneal = fibular.
n, nerve.

INJURY

SURAL NEUROPATHY

- *General*:
  - This is a pure sensory nerve.
- *Etiology*:
  - The nerve can be injured by compression from tight socks, a Baker's cyst or ganglion cyst, or trauma (e.g., laceration; Bryan et al., 1999).
- *Clinical presentation*:
  - This is a pure sensory nerve with no muscle involvement. The patient may complain of abnormal sensations to the lateral calf and foot. A positive Tinel's sign may be elicited along the course of the nerve.
- *EDX findings*:
  - **NCS:**
    - Abnormal SNAP. CMAP not applicable.
    - Note: Decreased or absent sural responses can be seen as a normal finding in elderly patients (Tavee et al., 2013).
  - **EMG:** Not applicable
- *Treatment*:
  - Interventions may include rehabilitation or surgical procedure.

SUPERIOR AND INFERIOR GLUTEAL NERVES

Anatomy

SUPERIOR GLUTEAL NERVE

- **Origin:**
  - These nerve fibers originate from the L4, L5, and S1 spinal nerve roots.

- **Course:**
  - In the pelvic region, the nerve passes through the sciatic notch superior to the piriformis muscle to innervate:
    - Gluteus medius
    - Gluteus minimus
    - Tensor fascia lata (TFL)

INFERIOR GLUTEAL NERVE

- **Origin:**
  - These nerve fibers originate from the L5–S2 spinal nerve roots.
- **Course:**
  - In the pelvic region, the nerve supplies the gluteus maximus.

Injury

- *General*:
  - These nerves are typically injured by iatrogenic causes, such as hip joint replacement, improper intramuscular injection, or pelvic masses.
- *Clinical presentation*:
  - A *superior gluteal neuropathy* presents with weakness of hip abduction and external rotation. A Trendelenburg gait demonstrating a pelvic tilt to the strong side will be seen.
  - In contrast, an *inferior gluteal neuropathy* presents with weakness of hip extension. Sensation is spared.
- *EDX findings*:
  - **NCS:** No SNAP or CMAP studies exist.
  - **EMG:**
    - *Superior gluteal neuropathy*: Abnormal findings in the gluteus medius, gluteus minimus, and TFL only
    - *Inferior gluteal neuropathy*: Abnormal findings in the gluteus maximus only

MONONEURITIS MULTIPLEX

- *General*:
  - Also known as mononeuropathy multiplex or multifocal neuropathy.
  - This is a type of multifocal peripheral neuropathy in which nerve damage occurs in two or more different nerve areas.
  - The condition refers to a group of symptoms (syndrome) rather than a disease. It can encompass a constellation of symptoms that include numbness, paresthesias, focal weakness or paralysis, and bowel/bladder dysfunction.
- *Etiology*:
  - Axonal injury can be caused by multiple etiologies, including inflammation (vasculitis), vascular compromise (occlusion), compression, and infection.
  - It is associated with a number of conditions:
    - Rheumatologic (polyarteritis nodosa, RA, systemic lupus erythematosus [SLE], scleroderma, Wegener's granulomatosis, and Sjrögren's)
    - Diabetes mellitus—most common; amyloidosis
    - Demyelinating (acute inflammatory demyelinating polyneuropathy [AIPD], chronic inflammatory demyelinating polyneuropathy [CIPD])
    - Infections (Lyme, leprosy, HIV, and hepatitis A, B, C)
    - Cancer (neurofibroma, malignant invasion)
- *Clinical presentation*:
  - Based on etiology and which peripheral nerves, cranial nerves, nerve roots, or plexus locations are involved.
  - Onset can be acute, subacute, or insidious presentation and pathology is asymmetric.

  - Sensation becomes abnormal and primarily affects pain and temperature (small fiber nerves) in a poorly localized pattern along with migratory arthralgias and myalgias.
  - Foot drop or give-way weakness can develop in an asymmetric distribution, and muscle stretch response (MSR) will be absent.
  - Abnormal findings on studies are based on etiology, but nerve infarctions cause primarily axonal degeneration and less commonly segmental demyelination. This will affect recovery, with axonal involvement being more prolonged and incomplete.
  - Nerve biopsy may provide definitive diagnosis.
- *EDX findings*:
  - EDX studies will demonstrate a typical pattern but are dependent on which nerve is involved.

EDX Findings in Mononeuritis Multiplex

MINIMAL DEMYELINATION FEATURES	AXONAL DEGENERATION
EDX Findings: Normal CV Increased in chronic conditions Normal temporal dispersion No conduction block	*EDX Findings*: Decreased amplitudes EMG abnormalities of spontaneous activity with decreased recruitment and polyphasic potentials of increased duration and amplitude, demonstrating collateral sprouting

CV, conduction velocity; EDX, electrodiagnostic; EMG, electromyography.

PERIPHERAL POLYNEUROPATHY (PERIPHERAL NEUROPATHY)

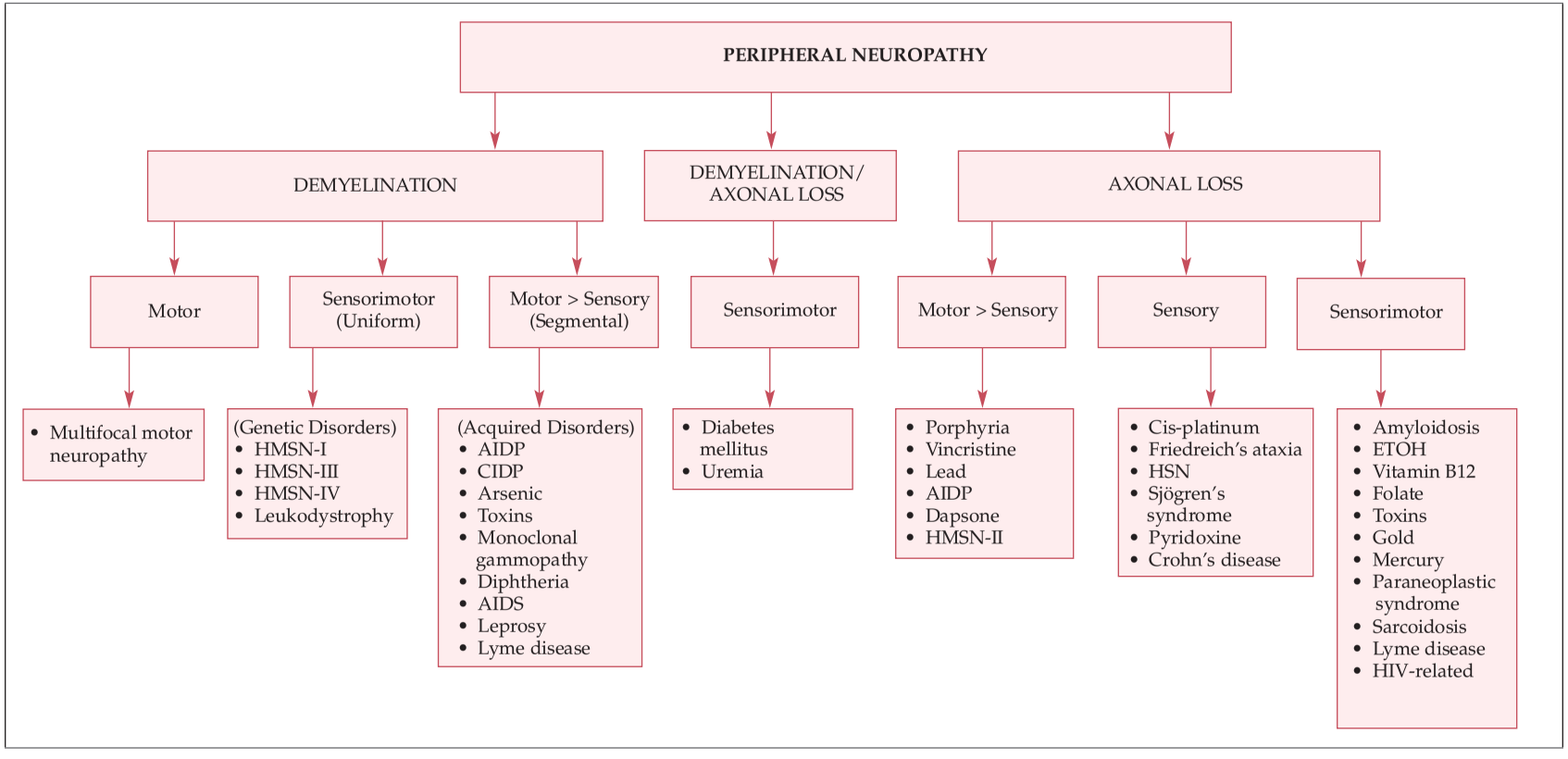
- Peripheral neuropathies (polyneuropathies) have numerous causes and are typically categorized as either inherited or acquired in origin.
- They can present with acute clinical symptoms or demonstrate a more insidious progression.
- EDX findings can give further insight on not only the type of neuropathy but also the nerve fibers affected to provide a greater understanding of the disorder.
- Peripheral neuropathies can affect the myelin and/or axons of the peripheral nerves. If demyelination becomes extensive, it can be accompanied by mild axonal damage.
- Determining a primary and predominating process is helpful in understanding the course of the disorder. The primary process can help determine the etiology of a neuropathy, while the predominant process can help assess the prognosis. They can be described as diffuse or multifocal.

PATTERN	PRESENTATION
Diffuse	Essentially involves all nerves in a length-dependent fashion to a relatively equal extent
Multifocal	Involves one or multiple nerves in an asymmetric or patchy distribution

ETIOLOGY (TABLES 5–33 AND 5–34)

- Symptom onset and location, presentation patterns, family history, and comorbidities are important to investigate to determine whether the condition is inherited or acquired.
- Certain clinical and EDX criteria are used to specifically classify the type of polyneuropathy, outlined on:
  - Table 5–33: Classification I
  - Table 5–34: Classification II

TABLE 5–33 Classification I



AIDP, acute inflammatory demyelinating polyneuropathy; CIDP, chronic inflammatory demyelinating polyneuropathy; ETOH, ethyl alcohol; HMSN, hereditary motor and sensory neuropathies; HSN, hereditary sensory neuropathy.

TABLE 5–34 Classification II

Diffuse axonal polyneuropathy	Toxins—Heavy metals; Drugs—Vincristine, alcohol Deficiency—Vitamin B6 deficiency Metabolic—Uremia, diabetes; paraneoplastic syndrome Hereditary—HMSN-II; Infectious—Lyme's disease, HIV
Multifocal axonal neuropathy	Microangiopathic—Vasculitis, diabetes; amyloidosis; paraneoplastic syndrome Infectious—CMV Metabolic—Porphyria; compression
Diffuse demyelinating polyneuropathy	Hereditary—HMSN-I, IV Deficiency—Hypothyroidism Toxic—Amiodarone, arsenic
Multifocal demyelinating neuropathy	Autoimmune—AIDP, CIDP; multiple compressions; leprosy

AIDP, acute inflammatory demyelinating polyneuropathy; CIDP, chronic inflammatory demyelinating polyneuropathy; CMV, cytomegalovirus; HMSN, hereditary motor and sensory neuropathies.

GENERAL OVERVIEW

Inherited Versus Acquired Peripheral Neuropathies

INHERITED PERIPHERAL NEUROPATHIES

- This is the largest group of progressive disorders that affect the PNS. Genetic testing plays a great role in confirming the diagnosis. These are typically categorized as:
  - Hereditary motor and sensory neuropathies (HMSN)
  - Hereditary sensory and autonomic neuropathies (HSAN)
  - Hereditary motor neuropathies (HMN)
- Classification of inherited polyneuropathies:
  - Primary classification is based on the presentation (HMSN, HSAN, and HMN).
  - Subclassification is based on inheritance pattern and EDX findings (demyelinating vs. axonal).
  - Specific gene mutation is also relevant.
- HMSN, also known as Charcot–Marie–Tooth disease, is the most commonly inherited peripheral polyneuropathy and is due to a duplication mutation of the PMP-22 gene.
  - A mutation related to the PMP-22 gene is also implicated as hereditary neuropathy with liability to pressure palsy (HNPP). This is due to a reciprocal deletion of the gene, causing this unique diagnosis, which presents with recurrent, short-term episodes of motor and sensory mononeuropathies. They can last from hours to a few weeks and are typically noted in regions of entrapments or from mild trauma.
- HSAN disorders are rare and primarily affect sensory and autonomic functions. There are several subclassifications with distinguishing features causing sensory loss, including pain and temperature, along with thermal dysregulation, bladder dysfunction, and cognitive deficits.
- HMN is characterized by progressive atrophy and weakness of the upper and lower extremities. This arises from abnormalities in the anterior horn cells and causes atrophy of the distal spinal muscles. They can progress to vocal cord and facial muscle paralysis.

ACQUIRED NEUROPATHIES

- These are associated with progressive medical conditions, inflammatory processes, or iatrogenic influences, most commonly related to:
  - Diabetes mellitus (most common)
  - Acute inflammatory demyelinating polyradiculopathy (AIDP)
  - Medications

- Acquired polyneuropathies are what we identify with most commonly due to its typical distal onset of symptoms.
  - This is seen because of the length-dependent manner in which it affects the axons of the nerve. It affects the distal-most part of the nerve and progresses proximally, as seen with diabetes mellitus, thyroid disorders, alcohol abuse, vitamin B12 deficiency, and critical illness neuropathy.
- Demyelinating inflammatory neuropathies can manifest as a polyradiculopathy, involving the peripheral nerves, the nerve roots, and the cranial nerves.
- Multiple nerves can also be affected in an asymmetric pattern manifesting either simultaneous neuropathies or randomly throughout the body or limb.
  - These findings may be seen in ischemic processes such as vasculitic neuropathies.
- Though less common, the upper limbs may demonstrate abnormalities first.
  - This can occur in vasculitic neuropathies, chronic inflammatory demyelinating polyradiculopathy (CIDP), HNPP toxicity, porphyria, inflammatory neuropathies, diabetes mellitus, and so forth.

CLINICAL PRESENTATION OF POLYNEUROPATHIES

- A classic triad has been described presenting more in the lower limbs than the upper limbs:
  - Sensory changes in a stocking/glove distribution
  - Distal weakness
  - Diminished/absent MSR
- However, inherited polyneuropathies typically present with:
  - Sensory loss
  - Ataxia
  - Increased incidence of muscle cramping
- The acquired polyneuropathies tend to demonstrate:
  - Burning
  - Pain
  - Paresthesias

ELECTRODIAGNOSTIC FINDINGS

The EDX criteria for peripheral neuropathy include evaluation of at least three limbs. Abnormalities of SNAPs, CMAPs, and MUAPs are dependent on the type and timing of the pathology affecting a nerve.

Basic Electrodiagnostic Findings in Peripheral Neuropathy

- Classic findings for a demyelinating versus axonal process are listed in the following chart. Conditions such as diabetes mellitus and uremia can present as a mixed pattern.
- Understanding the different findings seen in a hereditary versus acquired neuropathy aids in monitoring the diagnosis and can guide treatment options and prognosis.
- EDX abnormalities can include:
  - Prolonged latencies and increased CVs
  - Reduced or absent amplitudes
  - Prolonged or absent F-waves
  - Decreased persistence (“missing” waveforms on repetitive stimulations)
  - Increased chronodispersion (large latency difference between the fastest and slowest waveform on repetitive stimulation)

	DEMYELINATING INJURY	AXONAL INJURY
NCS	Prolonged distal latency Slowed conduction velocity Conduction block Increased temporal dispersion	Near-normal distal latency Near-normal conduction velocity Reduced SNAP/CMAP amplitude
EMG	No fibrillation potentials or positive sharp waves Myokymic discharge Decreased recruitment	(+) Fibrillation potentials (+) Positive sharp waves Decreased recruitment Increased duration and amplitude Polyphasic potentials

CMAP, compound muscle action potential; EMG, electromyography; NCS, nerve conduction studies; SNAP, sensory nerve action potential.

NCS FINDINGS	ACQUIRED NEUROPATHY	HEREDITARY NEUROPATHY
Conduction block	Positive	Negative
Conduction velocity	Focal slowing	Diffuse slowing
Temporal dispersion	Increased	Normal

NCS, nerve conduction studies.

SPECIAL STUDIES

- Small nerve fiber abnormalities and associated autonomic dysfunction may not be seen with conventional NCS and can require additional tests. These may be considered in patients with systemic symptoms such as orthostatic blood pressure changes, dry scaly skin, dry eyes, dry mouth, and so forth.
- Furthermore, small fiber neuropathies manifest as burning pain in the extremities with abnormal pinprick sensation but demonstrate normal standard NCS.

Autonomic Nerve Studies

- 🕮 **Norepinephrine synthesis and release:**
  - Norepinephrine is the primary neurotransmitter for postganglionic sympathetic adrenergic nerves. It is synthesized inside the nerve axon, stored within vesicles, then released by the nerve when an AP travels down the nerve. Following are the details for release and synthesis of norepinephrine:
    - The amino acid tyrosine is transported into the sympathetic nerve axon.
    - Tyrosine is converted to dihydroxyphenylalanine (DOPA) by tyrosine hydroxylase.
    - DOPA is converted to dopamine by DOPA decarboxylase.
    - Dopamine is transported into vesicles then converted to norepinephrine by dopamine b-hydroxylase.
    - An AP traveling down the axon depolarizes the membrane and causes calcium to enter the axon.
    - Increased intracellular calcium causes the vesicles to migrate to the axonal membrane and fuse with the membrane, which permits the norepinephrine to diffuse out of the vesicle into the extracellular space.
    - The norepinephrine binds to the postjunctional receptor and stimulates the effector organ response.
- 🕮 **Sympathetic skin response:**
  - The sympathetic skin response is a means of evaluating the unmyelinated, sympathetic nerve fibers of the PNS.

  - For median nerve testing using standard electrodes, E-1 can be placed on the palm and E-2 on the dorsum of the hand. The median nerve is stimulated at the wrist and elbow at the usual locations. Stimulation occurs over several minutes, and irregular stimulus intervals are required to prevent nerve habituation.
  - The stimulus sources used are electrical, coughing, noises, breathing, or tactile.
    - Current: 10 to 20 mA with a pulse width of 0.1 ms
    - Sweep speed: 500 ms/cm
    - LFF: 0.5 Hz
    - HFF: 2,000 Hz
    - Upper extremity CV: 1.57 ± 0.11 m/s
    - Lower extremity CV: 1.02 ± 0.07 m/s
- **Sinus arrhythmia:**
  - This cardiovagal innervation study is dependent on the normal heart rate variations that occur with respiration via parasympathetic activity.
  - Loss of this sinus arrhythmia represents a denervation process.
  - The test consists of measuring the R-R ratio with an EKG machine attached to the amplifier of the EMG.
- **Valsalva ratio:**
  - During a Valsalva maneuver, the heart rate varies in response to changes in blood pressure and intrathoracic pressure via sympathetic and parasympathetic activity. Four phases are measured using a standard EMG machine. The findings measured in Phase 2 and Phase 4 are used for monitoring. Phase 2 should demonstrate a heart rate increase, while Phase 4 should demonstrate a decrease in the normal population.
- 🕮 **Anal sphincter activity:**
  - EMG recording of the external anal sphincter has continuous activity at rest. There is a brief contraction in response to rapid rectal distension, and a preserved or increased activity during a prolonged substantial rectal distension during defecation in healthy adults.
- **Additional studies:**
  - Other studies can support the EDX findings when evaluating for peripheral neuropathies. These can include:
    - Nerve biopsies (inflammatory neuropathy, mononeuropathy multiplex, amyloidosis)
    - Skin biopsies (small fiber neuropathy)
    - Laboratory studies (blood glucose, B12 levels, serum protein immunofixation electrophoresis)
    - Autonomic testing

The following tables outline pertinent peripheral neuropatterns as they are defined by criteria in Classification I. Please refer to **Table 5–33** as an overview for **Tables 5–35 to 5–42**.

DIFFERENTIAL DIAGNOSIS OF FOOT DROP

- Diffuse polyneuropathy: Diabetes
- 🕮 Mononeuropathy: Common peroneal (fibular); deep peroneal (fibular) nerve and sciatic nerve
  - Lesions associated with axonal loss in the deep peroneal (fibular) nerves will result in EMG abnormalities in TA and EHL muscles.
  - Lesions associated with axonal loss in the common peroneal (fibular) nerve will result in EMG abnormalities in TA, EHL, and peroneus (fibularis) longus muscles.
  - Lesions associated with axonal loss in the sciatic nerve will result in EMG abnormalities in TA, EHL, peroneus (fibularis) longus, TP, FDL, and the short head of the biceps femoris muscles.

- Plexopathy
- Radiculopathy: L4–L5
- Central: Tumor, cerebrovascular accident (CVA), arteriovenous malformation (AVM), and spinal cord injury (SCI)

TABLE 5–35 Uniform Demyelinating Mixed Sensorimotor Neuropathies: Common Disorders

DISEASE	HMSN-I: CHARCOT–MARIE–TOOTH (CMT)	HMSN-III: DÉJÉRINE-SOTTAS	HMSN-IV: REFSUM'S DISEASE
Etiology	Autosomal dominant	Autosomal recessive	Autosomal recessive
Onset	Early childhood in the first 2 years	Birth–infancy	Approximately third decade
Clinical presentation	• Slowly progressive distal motor more than sensory abnormalities • Sensory loss in the lower limbs > the upper limbs • Abnormal vibration and proprioception • Stocking/glove pattern • Distal > proximal weakness • Abnormal MSR • Predominantly affects the intrinsic foot and lower leg anterior compartment musculature: Pes cavus and hammer toes • Bilateral foot drop: Steppage gait • Stork leg/champagne bottle leg appearance • Hypertrophy of peripheral nerves (greater auricular nerve) • Roussy–Levy syndrome: CMT associated with an essential tremor	• Severe progression • Sensory loss • Weakness • Abnormal MSR • Hypotonic/floppy baby • Delayed milestones • Ataxia • Pes cavus • Kyphoscoliosis • Nystagmus • Deafness	• Weakness • Abnormal MSR • Lower extremity wasting • Steppage gait • Ataxia • Retinitis pigmentosa (night blindness) • Cerebellar dysfunction • Deafness • Cardiac abnormalities • Cataract
Labs	CSF: Increased protein N Bx: Onion bulb formation from focal demyelination, then remyelination	CSF: Increased protein	CSF: Increased protein N Bx: Onion bulb formation Blood: High phytanic acid
EDX findings	NCS • SNAP: Abnormal • 🕮 CMAP: Abnormal, CV decreased 70%. No temporal dispersion or conduction block. EMG: Normal	NCS • SNAP: Abnormal • CMAP: Abnormal, CV is <6 m/s, latency is 7× slower than normal EMG: Normal	NCS • SNAP: Abnormal • CMAP: Abnormal, CV is 10 m/s EMG: Normal
Treatment	Rehabilitation; orthotics	Rehabilitation	Rehabilitation; phytanic acid absent diet

CMAP, compound motor action potential; CSF, cerebrospinal fluid; CV, conduction velocity; EDX, electrodiagnostic; EMG, electromyography; HMSN, hereditary motor sensory neuropathy; MSR, muscle stretch response; N Bx, nerve biopsy; NCS, nerve conduction study; SNAP, sensory nerve action potential.

TABLE 5–36 Segmental Demyelinating Motor and Sensory Neuropathies: Common Disorders

DISEASE	AIDP, GBS	CIDP	LEPROSY (HANSEN'S DISEASE)
Etiology	Possible viral attack on the myelin and Schwann cells	Possible immune-mediated response	*Mycobacterium leprae*
Onset	1–4 weeks post illness, vaccination, or surgery	Any age, peaks at 50–60 years of age	Immune status-dependent
Clinical presentation	• Male > female • 🕮 Ascending sensory abnormalities (ascending numbness is often the first sign) • Ascending symmetric weakness • Abnormal MSR • Possible respiratory and autonomic failure • Possibly bedridden within 2 days • CN involvement (Most common: CN VII affected, CN I and II unaffected) • Variants: Miller-Fisher syndrome, pure sensory	• Relapsing and remitting course • Sensory abnormalities • Symmetric weakness: Proximal > distal • Abnormal MSR • Less cranial nerve involvement	• Most common neuropathy worldwide • Sensory abnormalities • Wrist drop • Foot drop • Facial palsy
Labs	CSF: Increased protein, few mononuclear cells	CSF: Increased protein	N Bx: Foamy histiocyte invasion
EDX findings	NCS • SNAP: Abnormal • CMAP: Abnormal, temporal dispersion and conduction block: F-wave: Abnormal—first EDX sign EMG: Normal 🕮 Poorer prognosis if: NCS • CMAP: Amplitude <20% of normal, NCV <40% of normal • F-wave: Absent EMG: Abnormal activity (axonal involvement)	NCS • SNAP: Abnormal • CMAP: Abnormal; increased TD • F-wave: Abnormal EMG: Abnormal, if severe	NCS • SNAP: Abnormal • CMAP: Abnormal EMG: Abnormal, if severe
Treatment	Rehabilitation, plasmapheresis, IV immunoglobulins, steroids ineffective, respiratory support Majority of patients have near-complete recovery with only mild permanent sequelae within 3–6 months.	Rehabilitation, IV immunoglobulin, plasmapheresis, high-dose steroids	Rehabilitation, antileprosy treatment

AIDP, acute inflammatory demyelinating polyneuropathy; CIDP, chronic inflammatory demyelinating polyneuropathy; CMAP, compound motor action potential; CN, cranial nerve; CSF, cerebrospinal fluid; EDX, electrodiagnostic; EMG, electromyography; GBS, Guillain-Barré syndrome; IV, intravenous; MSR, muscle stretch response; N Bx, nerve biopsy, NCS, nerve conduction study; NCV, nerve conduction velocity; SNAP, sensory nerve action potential; TD, temporal dispersion.

TABLE 5–37 Axonal Motor > Sensory Neuropathies: Common Disorders

DISEASE	PORPHYRIA	TOXINS			AIDP AXONAL	HMSN-II CMT-II
Etiology	Defect heme synthesis	*Lead*	*Vincristine* Chemotherapy	*Dapsone* Leprosy treatment	Same as demyelination	Autosomal dominant
Clinical presentation	• Female > male • Lower limb pain • Limb weakness • Back and abdominal pain • Seizures • Mental status changes Reaction to medication, e.g., barbiturates, sulfonamides	• Progressive onset of upper-limb weakness • Radial neuropathy: Wrist drop (adult, child) • Encephalopathy (child) • Abdominal discomfort • Blue lines in the gums • Blindness • Epilepsy	• Lower limb paresthesias • Lower limb weakness • Abnormal MSR	• Ascending foot and hand neuropathy; side effects include methemoglobinemia	• Areflexia • Autonomic and cranial nerve involvement • Poorer prognosis than with pure demyelination • Associated with CMV and *Campylobacter jejuni* infection	• Onset commonly in the second decade • Weakness • Abnormal MSR • Less foot intrinsic involvement • Tremor • Ataxia
Labs	Urine: Deep red	Blood/urine: Lead basophilic stippling in RBCs, x-ray lead lines			CSF: Increased protein	N Bx: No onion bulb formation
EDX findings	NCS • SNAP: Abnormal • CMAP: Abnormal EMG • Abnormal	NCS • SNAP: Normal • CMAP: Abnormal EMG • Abnormal radial muscles	NCS • SNAP: Abnormal • CMAP: Abnormal EMG • Abnormal	NCS • SNAP: Normal • CMAP: Abnormal EMG • Abnormal	NCS • SNAP: Abnormal • CMAP: Abnormal EMG • Abnormal	NCS • SNAP: Abnormal • CMAP: Abnormal with preserved CV EMG • Abnormal paraspinal muscles
Treatment	Rehabilitation	Rehabilitation: Penicillamine, EDTA	Rehabilitation	Rehabilitation	Rehabilitation	Rehabilitation

AIDP, acute inflammatory demyelinating polyneuropathy; CMAP, compound muscle action potential; CMT, Charcot-Marie-Tooth; CMV, cytomegalovirus; CSF, cerebrospinal fluid; CV, conduction velocity; EDTA, ethylenediaminetetraacetic acid; EDX, electrodiagnostic; EMG, electromyography; HMSN, hereditary motor sensory neuropathy; MSR, muscle stretch response; NCS, nerve conduction study; N Bx, nerve biopsy; RBCs, red blood cells; SNAP, sensory nerve action potential.

TABLE 5–38 Axonal Sensory Neuropathies: Common Disorders

DISEASE	TOXINS	FRIEDREICH'S ATAXIA	SJÖGREN'S SYNDROME	TOXINS
Etiology	Cis-platinum	Autosomal recessive	Autoimmune disorder	Pyridoxine (B6)
Clinical presentation	• Painful paresthesias in the hands and feet • Abnormal sensation • Side effects: – Nephrotoxicity – Ototoxicity – Myelosuppression – GI complaints	• Onset: 2–16 years old • Abnormal sensation • Weakness • Abnormal MSR • Ataxia: Limb and trunk • Optic atrophy • Kyphoscoliosis • Dysarthria • Pes cavus deformity • Cardiomyopathy • Wheelchair use by 16 years of age	• Dry eyes • Dry mouth • Keratoconjunctivitis associated with rheumatoid arthritis • Gland involvement: – Parotid – Lacrimal – Salivary	• Abnormal sensation • Gait disturbances • Positive Lhermitte's sign • This may occur with doses of B6 >600 mg/day • Symptoms improve with drug withdrawal
Lab	N Bx: Abnormal large axons	N Bx: Abnormal large axons	N Bx: Abnormal large axons	N Bx: Abnormal large and small axons
EDX findings	NCS • SNAP: Abnormal • CMAP: Normal EMG • Normal	NCS • SNAP: Abnormal • CMAP: Normal EMG • Abnormal activity (motor unit remodeling)	NCS • SNAP: Abnormal • CMAP: Normal (can be abnormal) EMG • Abnormal (muscle remodeling)	NCS • SNAP: Abnormal • CMAP: Normal EMG • Abnormal: occ. fibs and pos. sharp waves
Treatment	Drug cessation	Rehabilitation	Rehabilitation	Stop vitamin B6

CMAP, compound motor action potential; EMG, electromyography; EDX, electrodiagnostic; GI, gastrointestinal; MSR, muscle stretch response; N Bx, nerve biopsy; NCS, nerve conduction studies; occ. fibs and pos., occasional fibrillations and positive; SNAP, sensory nerve action potential.

TABLE 5–39 Axonal Sensorimotor Neuropathies: Common Disorders

DISEASE	ETOH	AMYLOIDOSIS	SARCOIDOSIS
Etiology	Malnutrition or direct nerve injury	Amyloid deposition in DRG	Granulomatous disorder
Clinical presentation	• Sensory abnormalities • Foot or wrist drop • Muscle spasms • Korsakoff's psychosis • Wernicke's encephalopathy ± associated with a myopathy	• Sensory abnormalities • Weight loss • Ankle edema • Hepatomegaly • Purpura • Nephrotic syndrome • Congestive heart failure	• Low birth weight • Fatigue • Bilateral hilar adenopathy • Uveitis • Cranial nerve involvement (CN VII most common)
Labs	N Bx: Wallerian degeneration	Tissue Bx: (+) birefringence with Congo red staining	Blood: Increased ESR N Bx: Sarcoid tubercles
EDX findings	NCS • SNAP: Abnormal • CMAP: Abnormal EMG • Abnormal activity	NCS • SNAP: Abnormal • CMAP: Abnormal EMG • Abnormal activity	NCS • SNAP: Abnormal • CMAP: Abnormal EMG • Abnormal activity
Treatment	Vitamins, diet, stop alcohol consumption, orthotics	Rehabilitation	Rehabilitation

CMAP, compound motor action potential; CN, cranial nerve; DRG, dorsal root ganglion; EMG, electromyography; EDX, electrodiagnostic; ESR, erythrocyte sedimentation rate; ETOH, ethyl alcohol; N Bx, nerve biopsy; NCS, nerve conduction study; SNAP, sensory nerve action potential.

TABLE 5–40 Mixed Axonal and Demyelinating Neuropathies: Common Disorders

DISEASE	DIABETES MELLITUS	UREMIA
Clinical presentation	• Sensory abnormalities • Variants: Polyneuropathy, mononeuropathy, autonomic disorders, or amyotrophy • 🕮 Most common peripheral neuropathy in North America	• Occurs in 60% of patients with renal failure • Sensory abnormalities • Hypersensitivity to touch • Associated with restless leg syndrome
Labs	Blood: Elevated glucose N Bx: Small and large fiber abnormalities	Blood: Increased nitrogen and urea N Bx: Paranodal demyelination, axon loss
EDX findings	NCS • SNAP: Abnormal • CMAP: Abnormal EMG • Abnormal activity	NCS • SNAP: Abnormal • CMAP: Abnormal EMG • Abnormal activity
Treatment	Rehabilitation: Control blood sugar	Rehabilitation: Dialysis, kidney transplant

CMAP, compound motor action potential; EDX, electrodiagnostic; EMG, electromyography; N Bx, nerve biopsy; NCS, nerve conduction studies; SNAP, sensory nerve action potential.

TABLE 5–41 Demyelinating Motor Neuropathy

DISEASE	MULTIFOCAL MOTOR NEUROPATHY
Etiology	Immune-mediated disorder causing inflammatory demyelination and remyelination
Clinical presentation	Slowly progressing focal weakness Spreading fasciculations and cramps Atrophy and myokymia Asymmetric reduced MSR Sensation is normal Resembles MND
Labs	Nerve Bx: Endoneurial edema, lymphocytic inflammation, reduced myelin density, onion bulb formation (Findings resemble CIPD, except MMN only affects motor nerves.) Blood: Increased anti-GM1 antibody titers
EDX findings	• SNAP: Typically normal, though mild changes have been noted • CMAP: Latencies typically abnormal, amplitudes can be normal in weak muscles or show an 80% drop, decreased CV • MMN is defined by multifocal motor conduction block. More than one site of CB can occur in a single motor nerve. • F-wave: Abnormal • EMG: Abnormal spontaneous activity, including fasciculations and myokymic discharges
Other	Findings that help distinguish MMN from MND: • In MMN activity is confined to the muscles of clinical weakness. • In MND it is diffusely distributed. • In MMN activity can be traced back to peripheral nerve territories. • In MND it can be traced to a spinal segmental pattern.
Treatment	High-dose IVIg

Bx, biopsy; CB, conduction block; CIPD, chronic inflammatory demyelinating polyneuropathy; CMAP, compound motor action potential; CV, conduction velocity; EDX, electrodiagnostic; EMG, electromyography; IV, intravenous; IVIg, IV immunoglobulin; MND, motor neuron disease; MMN, multifocal motor neuropathy; MSR, muscle stretch response; SNAP, sensory nerve action potential.

TABLE 5–42 HIV-Related Neuropathies

FIVE MAJOR CATEGORIES
1. 🕮 *Distal Symmetric Polyneuropathy*: This is the most common type of neuropathy. It primarily affects sensory and autonomic fibers, with motor disruption occurring in advanced cases. Painful paresthesias begin in the toes followed by the fingers and advance proximally up the extremities.
2. *Inflammatory Demyelinating Polyneuropathy*: This presents in a similar manner as AIDP or CIDP. However, pleocytosis in the CSF with elevated protein distinguishes it from idiopathic AIDP/CIDP.
3. *Mononeuropathy Multiplex*: Thrombosis of the vasa nervorum leads to multiple lesions in various nerves. This causes primarily axonal loss with relative myelin sparing. This results in abnormal spontaneous activity on needle exam, but normal NCS with decreased amplitudes.
4. *Progressive Polyradiculopathy*: This results from cytomegalovirus causing severe asymmetrical pain, numbness, and motor deficits in the legs. Bowel and bladder dysfunctions along with impaired MSR are also noted.
5. *Autonomic Neuropathy*: A group of symptoms associated with damage of the nerves responsible for functions that regulate blood pressure, heart rate, bowel and bladder emptying, digestion, and so forth.
EDX findings: • NCS: Abnormal SNAPs and CMAPs • EMG: Abnormal activity • Most commonly presents with demyelination and axonal loss
Treatment: Rehabilitation, medications

AIDP, acute inflammatory demyelinating polyneuropathy; CIDP, chronic inflammatory demyelinating polyneuropathy; CMAP, compound motor action potential; CSF, cerebrospinal fluid; EDX, electrodiagnostic; EMG, electromyography; MSR, muscle stretch response; NCS, nerve conduction study; SNAP, sensory nerve action potential.

NEUROMUSCULAR JUNCTION DISORDERS

- These disorders hinder the production, release, or uptake of ACh at the NMJ. A low safety factor causes the amplitude of the EPPs to fall below the threshold needed to generate a muscle fiber AP. This occurs due to an alteration of quantal response or content (Table 5–43).
- **Myasthenia gravis** (MG) is a disorder resulting in a decreased quantal response due to an autoimmune response against postsynaptic ACh receptors. This leads to reduced MEPP amplitudes, but their frequency remains normal (quantal content is normal; Figure 5–119).
- **Lambert–Eaton myasthenic syndrome** (LEMS, myasthenic syndrome) is a disorder resulting in decreased quantal content leaving the presynaptic cleft, resulting in normal MEPP amplitudes but with decreased frequency (quantal response is normal).

ELECTRODIAGNOSTIC FINDINGS

- Evaluation of the NMJ involves typical NCS with EMG, as well as the addition of repetitive nerve stimulations (RNS) and single-fiber EMG (SFEMG; if needed).
- **NCS**:
  - SNAP: Typically normal. Occasionally, sensory neuropathies can be associated with a paraneoplastic syndrome seen with LEMS.
  - CMAP: Normal or decreased amplitude. If low, this should be followed by a 10-second maximum voluntary contraction, followed by a single stimulation. Increased amplitudes >100% compared with premaximum contraction are seen in LEMS.
- **EMG:** Can be normal or abnormal
  - In severe cases, abnormal spontaneous activity can be seen. Short-duration, low-amplitude MUAPs with early recruitment are associated with blocking.
  - MUAPs become unstable (variable amplitudes and configurations—Figure 5–120).
- RNS and SFEMG:
  - Abnormal RNS and SFEMG studies. See next section.

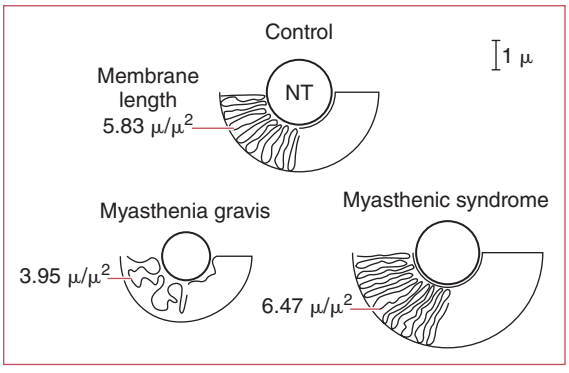


FIGURE 5–119 Postsynaptic membrane changes. MG–Simplification of the postsynaptic membrane. The NMJ demonstrates a reduction in the number of postsynaptic junctional folds. MS–Hypertrophy of postsynaptic membrane; the NMJs demonstrate an increase in the complexity of the postsynaptic membrane architecture.
MG, myasthenia gravis; MS, myasthenic syndrome; NMJ, neuromuscular junction.

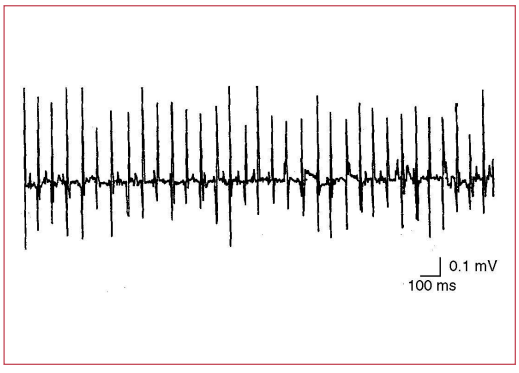


FIGURE 5–120 Unstable MUAP. Same MUAP with varying amplitudes. This is seen in patients with MG; amplitude variations are from neuromuscular junction blocking.
MG, myasthenia gravis; MUAP, motor unit action potential.

TABLE 5–43 Neuromuscular Junction Disorders

DISEASE	MYASTHENIA GRAVIS	LAMBERT–EATON SYNDROME (LEMS)	BOTULISM
Location	*Postsynaptic*	*Presynaptic*	*Presynaptic*
Etiology	• A disorder of neuromuscular transmission due to an autoimmune response in which polyclonal antibodies are directed against the Muscle Specific Tyrosine Kinase (MuSK) of the postsynaptic membrane. • Associated with thymic disorder or thymic tumor.	• A disorder of neuromuscular transmission due to an autoimmune response against the active sites (voltage-gated P/Q Ca channels on the presynaptic membrane). • This decreases Ca^{++} entry into the cell, causing a decreased release of ACh into the synaptic cleft. • Associated with small cell (oat cell) carcinoma of the lung (50% are paraneoplastic).	• A disorder of neuromuscular transmission caused by *Clostridium botulinum* toxins blocking presynaptic exocytosis of ACh from the nerve terminal. • Associated with ingestion of contaminated raw meat, fish, canned vegetables, and raw honey.
Onset	Bimodal distribution First Peak: 20–30 years, Female > male Second Peak: 60–80 years, Female = male	Bimodal distribution First Peak: 40 years, Female > male Second Peak: 60 years, Male > female	Begins 2–7 days after ingestion
Clinical presentation	• Painless proximal fatigue and weakness. Ocular weakness is the most common. • Exacerbated with exercise, heat, or time of day (evening) • Normal MSR • 📖 Facial or bulbar symptoms: – Ocular weakness (ptosis) – Diplopia – Dysphagia – Dysarthria • Improved with rest • Edrophonium (Tensilon) Test: 2-mg dose followed by an 8-mg dose, improvement begins in 1 minute	• Proximal fatigue and weakness • Mainly affects the lower limbs first (quadriceps) • Abnormal MSR • Exacerbated with rest • Improved with exercise • Viselike grip • Rarely involves the neck, facial, or bulbar muscles in contrast to MG • Autonomic symptoms: – Dry mouth – Erectile dysfunction – Constipation	• Decreased deep tendon reflexes • Bulbar symptoms are noted first: – Ocular weakness (ptosis) – Dysphagia – Dysarthria • GI symptoms: – Diarrhea, N/V – Widespread paralysis or flaccidity • Abnormal MSR • Respiratory and cardiac dysfunction
Labs	Muscle biopsy: Simplification of the postjunctional membrane with loss of junctional folds and receptors (see Figure 5–119) Antibody testing: • Anti-ACh receptor antibodies • Anti-MuSK antibodies	Muscle biopsy: Overdevelopment of neuromuscular junction (see Figure 5–119) Decreased active zones are noted Antibodies against voltage-gated Ca^{+} channels Monitor for cancer	Botulinum toxin: Found in stool or blood serum

TABLE 5–43 Neuromuscular Junction Disorders (*continued*)

DISEASE	MYASTHENIA GRAVIS	LAMBERT–EATON SYNDROME (LEMS)	BOTULISM
EDX findings	NCS • Normal SNAP and CMAP • >10% decrement on low rate rep. stim. EMG • Unstable MUAP, drop-off occurs with sustained contraction (see Figure 5–120) • See single-fiber EMG	NCS • SNAP: Normal • CMAP: Low amplitude • >10% decrement on low rate rep. stim. EMG • Unstable MUAP, drop-off occurs with sustained contraction • See single-fiber EMG	NCS • SNAP: Normal • CMAP: Abnormal amplitude • >10% decrement on rep. stim. study EMG • Unstable MUAP • See single-fiber EMG
Treatment	• Thymectomy • Anticholinesterase drugs: – Mestinon (Pyridostigmine) 30 mg q 4–6 hours • Corticosteroids • Immunosuppressive agents • Plasmapheresis • One-third improve spontaneously • IV immunoglobulin	• Treat malignancy • Corticosteroids • Immunosuppressive agents • Plasmapheresis • Guanidine: Increases ACh quanta – Side effects: GI, bone marrow suppression, renal tubular necrosis • 3,4-diaminopyridine • IV immunoglobulin	• Treat with trivalent ABE antitoxin in the first 24 hours • Supportive with intubation for respiratory failure • Recovery occurs from collateral sprouting

ACh, acetylcholine; CMAP, compound muscle action potential; EDX, electrodiagnostic; EMG, electromyography; GI, gastrointestinal; IV, intravenous; MG, myasthenia gravis; MSR, muscle stretch response; MUAP, motor unit action potential; NCS, nerve conduction study; N/V, nausea and vomiting; rep. stim., repetitive stimulation; SNAP, sensory nerve action potential.

REPETITIVE NERVE STIMULATION (FIGURE 5–121 AND TABLE 5–44)

- These are studies in which a repeated supramaximal stimulation of a motor nerve is performed.
- A series of CMAPs are recorded for pathologic amplitude changes. Muscles should be evaluated in a proximal progression if an abnormality is suspected but not demonstrated.
- The study is best performed on the clinically weak muscle(s). However, due to the ease of the examination, it is typically started in the hand intrinsics. If no abnormality is noted, then progression to more proximal muscles is performed.
- Proper setup is essential to obtain the appropriate responses. Prior to starting the study, cholinesterase inhibitors should be held for 12 hours, if medically cleared.

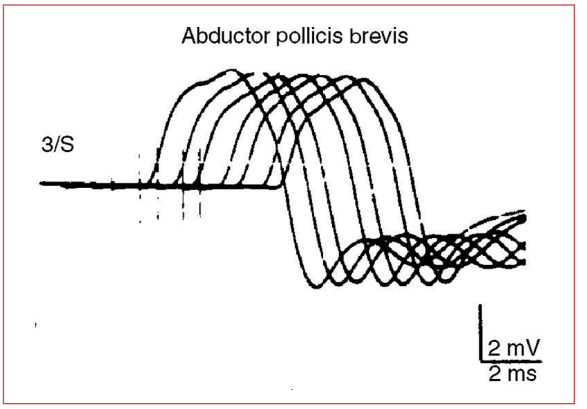


FIGURE 5–121 Repetitive nerve stimulation: Normal response.

TABLE 5–44 Muscle Evaluation for RNS

PROGRESSION	MUSCLES
First	ADM or APB
Second	Deltoid
Third	Trapezius
Fourth	Orbicularis oculi

ADM, abductor digiti minimi; APB, abductor pollicis brevis; RNS, repetitive nerve stimulation.

REPETITIVE NERVE STIMULATION SETUP

- Immobilize the electrode.
- Immobilize the limb.
- Stimulate at a supramaximal level.
- Optimize limb temperature (approximately 30°C).
- Minimize electrode gel.
- Stop anticholinesterase inhibitors.

🕮 ABNORMALITY

A >10% decrease in amplitude from the first to fifth waveform is significant for pathology.

Low-Rate Repetitive Stimulation (Figure 5–122)

- This repetitive stimulation test is performed at a rate of 2 to 3 Hz.
- Each stimulus causes the EPP amplitude to drop. If the safety factor is decreased, the potential will fall below the threshold necessary for activation. This results in a decrease of the MUAP amplitude (Table 5–45).

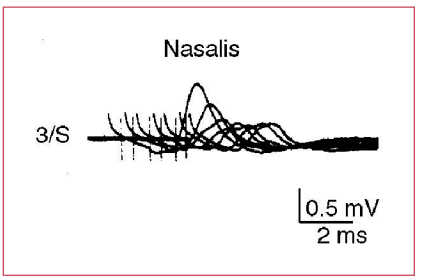


FIGURE 5–122 Low-rate repetitive stimulation decremental response.

- An abnormality is considered when a CMAP demonstrates >10% amplitude reduction between the first and fourth waveforms.
- An increase in waveform can be seen if more stimulations are provided due to mobilization of secondary ACh stores.
- A typical U-shaped decrement can be seen in MG.

TABLE 5–45 LRRS Amplitude Changes

DISORDER	AMPLITUDE CHANGE
Myasthenia gravis	>10% decrement
Lambert–Eaton syndrome	>10% decrement
Botulism	>10% decrement

LRRS, low-rate repetitive stimulation.

Postactivation Facilitation (PAF)

After a decrement is noted with LRRS, a 30- to 60-second isometric contraction or tetany-producing stimulation (50 Hz) should be performed. Postactivation Facilitation (PAF) demonstrates a repair in the CMAP amplitude with an immediate follow-up LRRS because of an improvement in neuromuscular transmission.

CMAP, compound muscle action potential; LRRS, low-rate repetitive stimulation.

Postactivation Exhaustion (PAE)

This response is seen as a CMAP amplitude decreases. It occurs with an LRRS performed every minute for 5 minutes after an initial 30- to 60-second isometric contraction. The greatest drop-off is between 2 and 4 minutes. This test should be used if a decrement does not present with the initial LRRS, but a diagnosis of an NMJ disorder is suspected **(Figure 5–123)**.

CMAP, compound muscle action potential; LRRS, low-rate repetitive stimulation; NMJ, neuromuscular junction.

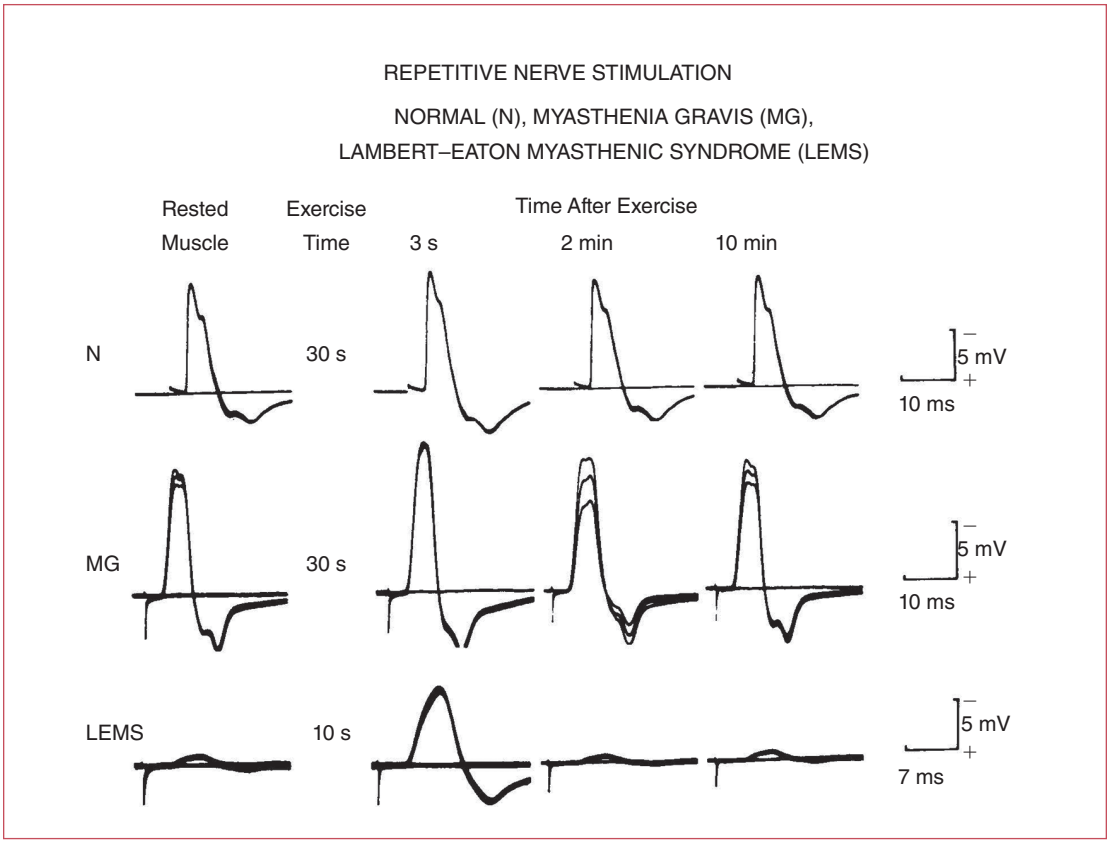


FIGURE 5–123 Repetitive stimulation (a decrement must be reproducible on a number of trials).

High-Rate Repetitive Stimulation (Figure 5–124, Table 5–46)

- This repetitive stimulation test is performed at a rate of 10 to 50 Hz. It causes an accumulation of calcium in the cell, which assists ACh release and repairs the waveforms.
- High-rate repetitive stimulation (HRRS) is uncomfortable and is typically performed if a patient is unable to perform a 30- to 60-second maximal isometric contraction.

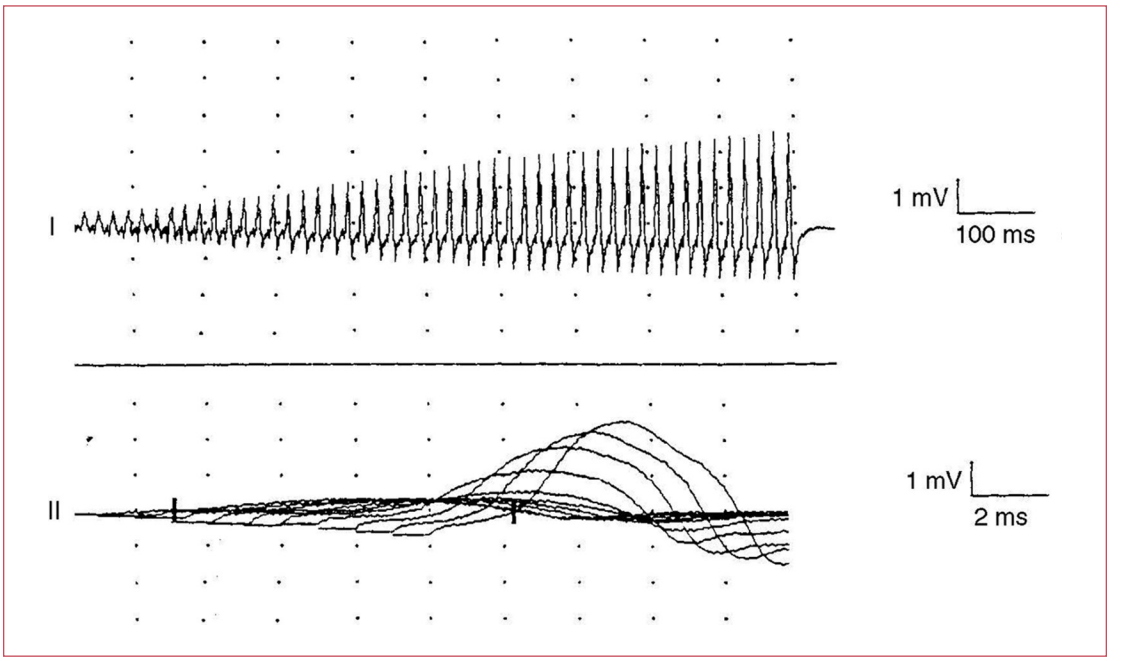


FIGURE 5–124 High-rate repetitive stimulation. (I) Increment with 50-Hz stimulation. (II) Increment with voluntary contraction (50-Hz simulation/train of 50, femoral/rectus femoris, 500% facilitation).

TABLE 5–46 High-Rate Repetitive Stimulation Amplitude Changes

DISORDER	AMPLITUDE CHANGE
Myasthenia gravis	Decrement demonstrated and partial rebounds
Lambert–Eaton syndrome	200%–300% increment
Botulism	Mild increment

Pseudofacilitation (Figure 5–125)

- This is a normal reaction and demonstrates a progressive increase in CMAP amplitude with HRRS or voluntary muscle contraction.
- It represents a decrease in temporal dispersion due to increased synchronicity of muscle fiber contraction. The waveforms produced maintain a constant area under the curve though the amplitude appears increased because the duration is decreased.

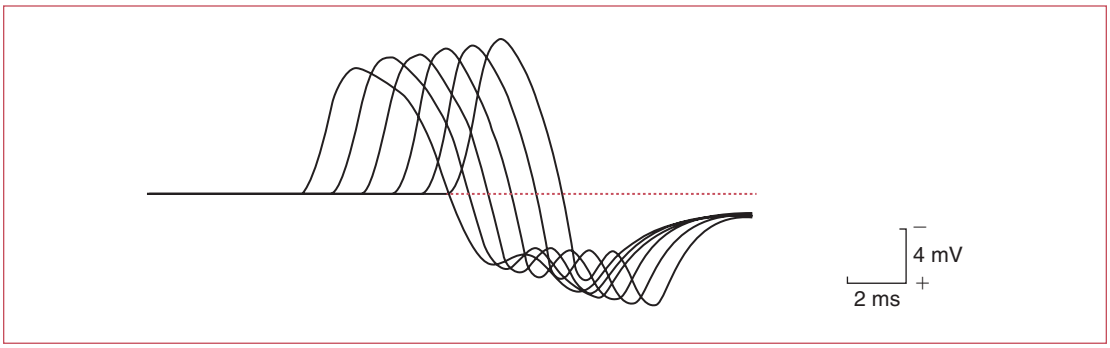


FIGURE 5–125 Pseudofacilitation. Repetitive nerve stimulation study in a normal subject. The successive M waves were recorded with surface electrodes over the hypothenar eminence (abductor digiti quinti) during ulnar nerve stimulation at a rate of 30 Hz. Pseudofacilitation may occur in normal subjects with repetitive nerve stimulation at high (20–50 Hz) rates or after strong volitional contraction, and probably reflects a reduction in the temporal dispersion of the summation of a constant number of muscle fiber action potentials due to increases in the propagation velocity of action potentials of muscle cells with repeated activation. Pseudofacilitation should be distinguished from facilitation. The recording shows an incrementing response characterized by an increase in the amplitude of the successive M waves with a corresponding decrease in the duration of the M wave resulting in no change in the area of the negative phase of the successive M waves.

RNS Findings in NMJ Disorders

	NMJ DISORDER		
TEST	**MYASTHENIA GRAVIS**	**LEMS**	**BOTULISM**
CMAP amplitude	Normal or reduced	Decreased	Decreased
RNS • LRRS	• >10% decrement noted between the first and fourth to fifth stimulation	• >10% decrement in amplitude	• >10% decrement in amplitude, or variable changes
• HRRS*	• Decrement and partial repair	• 200%–300% increment	• Mild increment
PAF	20%–50% improvement	>100% improvement	>40% improvement
PAE	Observed 2–4 minutes after maximal voluntary contraction	A train of five stimuli every minute for 5 minutes to monitor for decrease	Absent

CMAP, compound muscle action potential; HRRS, high-rate repetitive stimulation; LEMS, Lambert–Eaton myasthenic syndrome; LRRS, low-rate repetitive stimulation; NMJ, neuromuscular junction; PAE, postactivation exhaustion; PAF, post activation facilitation; RNS, repetitive nerve stimulation.
*HRRS is only done if patient cannot perform 30 to 60 seconds of maximal isometric contraction.

SINGLE-FIBER ELECTROMYOGRAPHY

- This is a study that monitors the parameters of single muscle fiber APs. It is useful if repetitive stimulation of at least three muscles is normal and an abnormal diagnosis is still suspected.
- SFEMG is the most sensitive test for NMJ disorders but has low specificity.
- Abnormalities can be associated with NMJ disorders, motor neuron disorders, and peripheral neuropathies.

PARAMETERS

- **Fiber density (Figure 5–126):**
  - This represents the number of single fibers belonging to the same motor unit within the recording radius of the electrode. The fiber density (FD) is determined by dividing the number of single muscle fiber APs at 20 sites by 20.
  - An FD of 1.5 is normal. Higher than this represents a denervation and reinnervation process.

- **Jitter (Figure 5–127):**
  - During voluntary contraction, a small variation exists between the interpotential discharges of two muscle fibers belonging to the same motor unit. This variation is normally 10 to 60 µs. It is typically considered abnormal if it is longer than this.
  - Disorders of neuromuscular transmission affect the safety factor and cause a delay in the time for an EPP to reach threshold for a muscle fiber AP, which increases the jitter between the two neighboring muscle fibers. Reinnervation through collateral sprouting after a nerve injury also can cause a delay. The immature NMJs have poor activation, resulting in increased jitter within the first month.
  - This is seen in conditions including amyotrophic lateral sclerosis (ALS), NMJ disorders, axonal neuropathies, and myopathies.
- **Blocking:**
  - This is an abnormality that occurs when a single muscle fiber AP fails to appear. It occurs if the jitter becomes >100 µs. It typically resolves in approximately 1 to 3 months, after reinnervation is completed. However, the increased jitter may take approximately 6 months to resolve.

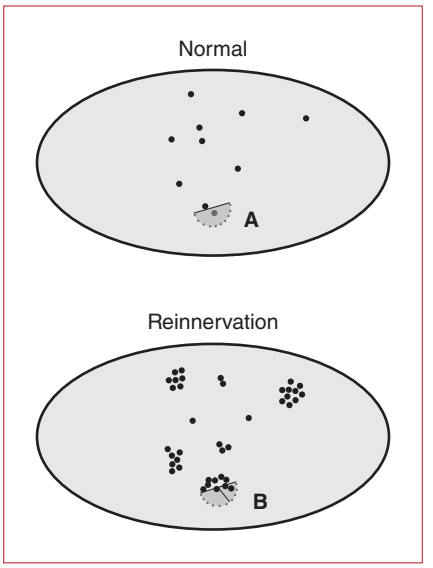


FIGURE 5–126 Increased fiber density. The dots represent single muscle fibers of one motor unit with the recording radius. (A) Normal muscle (action potentials from 1 to 2 fibers recorded). (B) Reinnervation (action potentials from many fibers recorded).

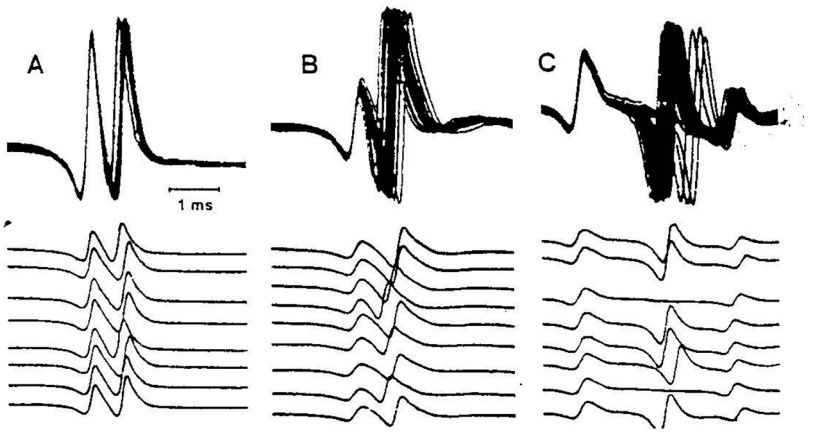


FIGURE 5–127 Single-fiber EMG recordings. *Top*: Superimposed view. *Bottom*: Rastered view. (A) Normal. (B) Increased jitter. (C) Increased jitter with blocking.
EMG, electromyography.

MYOPATHIES

- These are skeletal muscle fiber disorders that can occur from a variety of etiologies.
- Important factors to consider in its diagnosis include age of onset, developmental milestones, familial involvement, prodromal illness, and patient history.
- Currently genetic testing has demonstrated a greater ability to classify the type of myopathy.
- Please refer to Chapter 10, Pediatric Rehabilitation, for additional information on this topic.

ETIOLOGY (TABLE 5–47)

TABLE 5–47 Etiology of Myopathies

DYSTROPHIC	CONGENITAL	METABOLIC	INFLAMMATORY	ENDOCRINE	TOXIC	STEROID
• Duchenne • Becker • Limb-girdle disorder • Facioscapulohumeral • Myotonic	• Central core • Nemaline rod • Centronuclear • Fiber type disproportion	• Acid maltase deficiency • Myophosphorylase deficiency • Phosphofructokinase deficiency • Hyperkalemic periodic paralysis • Hypokalemic periodic paralysis	• Polymyositis/dermatomyositis • Sarcoidosis • Viral • Bacterial • Parasitic • Inclusion body myositis	• Thyroid parathyroid • Adrenal pituitary	• Alcohol • Diuretic • Vincristine • Steroid	• Corticosteroid use

The Role of Dystrophin

- Dystrophin is a protein found in the sarcolemma of normal muscle. It provides mechanical support and structural integrity for the muscle membrane cytoskeleton.
- Mutation in the dystrophin gene leads to muscle fiber necrosis. Patients present with clinical symptoms of myalgias, fatigue, and weakness.
- Muscle biopsies help differentiate between dystrophinopathies. In Duchenne muscular dystrophy, dystrophin is absent or markedly deficient. In Becker's muscular dystrophy, the abnormalities are less severe.

CLINICAL PRESENTATION

- The patient may demonstrate muscle-related changes presenting as atrophy, hypertrophy, abnormal MSR, weakness, hypotonia, gait abnormalities, or myotonia.
- Myotonia is a painless delayed relaxation of skeletal muscles following a voluntary contraction. It is exacerbated by cold but relieved with exercise, Dilantin, procainamide, and calcium channel blockers.
- Arthrogryposis, which is a fixed deformity of the extremities due to intrauterine hypomobility, may occur in newborns from myopathies, muscular dystrophies, or oligohydramnios. A hallmark sign of myopathy is the inability to generate a forceful contraction.

ELECTRODIAGNOSTIC FINDINGS

NCS

- SNAP: Normal
- CMAP: Decreased amplitude with significant muscle fiber atrophy. Normal latencies and CVs.

EMG

- Classic findings are low-amplitude, short-duration, polyphasic MUAP with early recruitment (Table 5–48).
- Resting activity: Abnormal activity depends on the type of disorder involved (Table 5–49).

Quantitative Electromyography

- This study may provide a more detailed measurement of the MUAPs. It is a better indication of waveform duration, which is a sensitive parameter for diagnosing myopathies. The mean duration is calculated using 20 MUAPs and on a screen set with a trigger and delay line. This avoids superimposing MUAPs and falsely creating a polyphasic.

TABLE 5–48 Recruitment: Early Onset With Minimal Effort

PRESENTATION	POSSIBLE CAUSES OF MUAP ALTERATIONS
SDSA	These classic polyphasic potentials are due to loss of muscle fibers.
LDLA	These polyphasic potentials are due to collateral sprouting.
Unstable	These variable amplitude potentials are due to blocking of immature NMJs, which are formed at the beginning of collateral sprouting.

LDLA, long-duration, large-amplitude; MUAP, motor unit action potential; NMJ, neuromuscular junction; SDSA, short-duration, small-amplitude.

TABLE 5–49 Abnormal Spontaneous Activity in Myopathies

FIBRILLATIONS AND POSITIVE SHARP WAVES	COMPLEX REPETITIVE DISCHARGE	MYOTONIC DISCHARGE
• Polymyositis • Dermatomyositis • Inclusion body myopathy • Trichinosis • Toxic myopathies • Direct muscle trauma • Rhabdomyolysis • Acid maltase deficiency • Myotubular myopathy • Hyperkalemic periodic paralysis • Nemaline rod • Sarcoid myopathy • Muscular dystrophies	• Polymyositis • Dermatomyositis • Muscular dystrophies • Schwartz-Jampel syndrome • Inclusion body myopathy	• Myotonia congenita • Myotonic dystrophy • Paramyotonia congenita • Hyperkalemic periodic paralysis • Acid maltase deficiency • Hypothyroid myopathy • Myotubular myopathy • Chloroquine myopathy • Diazo cholesterol intoxication • Polymyositis • Dermatomyositis

Repetitive Nerve Stimulation

- A normal or a decremental response can occur. This is due to the reduced safety factor found in regenerating immature NMJs that form during recovery or reinnervation.

Single-Fiber Electromyography

- This can demonstrate increased jitter, FD, and blocking.

Additional Testing: Muscle Biopsy

TYPE I FIBER ATROPHY	TYPE II FIBER ATROPHY
• Myotonic dystrophy • Nemaline rod myopathy • Fiber type disproportion	• Steroid myopathy • Myasthenia gravis • Deconditioning

TYPES OF MYOPATHIES

- The following tables outline pertinent myopathic patterns.
- Please refer to **Table 5–47** as an overview for **Tables 5–50** through **5–56**.

TABLE 5–50 Dystrophic Myopathies: Common Disorders

DISORDER	DUCHENNE MUSCULAR DYSTROPHY (DMD; MOST COMMON MYOPATHY)	BECKER MUSCULAR DYSTROPHY (BMD)	MYOTONIC DYSTROPHY (STEINERT'S; SECOND MOST COMMON)	FACIOSCAPULOHUMERAL DYSTROPHY (FSH OR FSHD)
Etiology	X-linked recessive (xp21), spontaneous	X-linked recessive	Autosomal dominant	Autosomal dominant
Onset	3–5 years old	Adulthood	Infant	Childhood–early adult
Course	Severely progressive (death by 20s)	Slowly progressive		Spreads to other muscles
Clinical presentation	• Proximal muscle weakness (pelvic girdle) • Abnormal MSR • Increased lumbar lordosis • Ambulation difficulties: Toe walking (<5 years), clumsy running (<7 years) • Gower's sign: Difficulty rising from the floor due to hip and knee extensor weakness • Calf pseudohypertrophy with fat and fibrous tissue • Contractures: Iliotibial band (first), Achilles tendon • Scoliosis, causing cardiomyopathy and restrictive lung disease • Possible mental retardation • Wheelchair by 12 years old • Extraocular muscles are spared	• Proximal weakness • Calf pseudohypertrophy • Cardiomyopathy • Less mental retardation than DMD	• Weakness: Distal > proximal myotonia with sustained grip • Hatchet face (wasting of the temporalis and masseter) • Frontal balding • Poor vision • Ptosis • Impotence • Hypertrichosis • Mental retardation • Cardiac abnormalities • Endocrine abnormalities • Congenital myotonic dystrophy: • "Shark mouth" appearance • Facial diplegia • Possible club foot	• Proximal muscle weakness • Facial droop • Weak eye closing • Weak forehead wrinkling • Arm atrophy with deltoid and forearm sparing (Popeye arm) • Cataracts (dry sclera) • Retinopathy • Lip protrusion • Transverse smile • Frontal balding • Testicular atrophy • Extraocular muscles are spared • #1 muscle to test in FSH is tibialis anterior • Inability to whistle
Labs	M Bx: No dystrophin, internal nuclei variation in fiber size Blood: Increased CPK and aldolase EKG: Abnormal	M Bx: Decreased dystrophin (15%–85%). Blood: Increased CPK	M Bx: Type I fiber atrophy with Type II hypertrophy. No dystrophin involvement.	M Bx: Scattered fiber necrosis and regeneration. Inflammatory infiltrate may be noted.
EDX findings	NCS • SNAP: Normal • CMAP: ± Decreased amplitude EMG • AA (rare), ER, ± SDSA MUAP	NCS • SNAP: Normal • CMAP: ± Decreased amplitude EMG • AA (rare), ER, SDSA MUAP	NCS • SNAP: Normal • CMAP: ± Decreased amplitude EMG • AA (rare), ER, SDSA MUAP, myotonia	NCS • SNAP: Normal • CMAP: Decreased amplitude in the involved muscles EMG • AA, ER, SDSA MUAP
Treatment	Rehabilitation. Scoliosis surgery before the vital capacity is below 35% (usually due to a curve of >30 degree). Prednisone.	Rehabilitation: Bracing. Tendon lengthening, possible scoliosis surgery.	Rehabilitation: Bracing. Medications: Procainamide, Dilantin, and quinine (PDQ). May need a pacemaker.	Rehabilitation

AA, abnormal activity; CMAP, compound muscle action potential; CPK, creatine phosphokinase; EDX, electrodiagnostic; EMG, electromyography; ER, early recruitment; M Bx, muscle biopsy; NCS, nerve conduction study; MSR, muscle stretch response; MUAP, motor unit action potential; SDSA, short-duration, small-amplitude; SNAP, sensory nerve action potential.

TABLE 5–51 Congenital Myopathies: Common Disorders

DISORDER	CENTRAL CORE DISEASE	NEMALINE ROD MYOPATHY	CENTRONUCLEAR MYOTUBULAR	FIBER TYPE DISPROPORTION
Genetic etiology	Autosomal dominant	Autosomal dominant/recessive	X-linked recessive	Variable
Onset	Infancy	Infancy	Infancy	Infancy
Clinical presentation	• Floppy infant/hypotonia • Proximal weakness • Congenital hip dislocation • Delayed milestones • 🕮 Associated with malignant hyperthermia	• Floppy infant/hypotonia • Diffuse weakness • Facial involvement • Narrowed long face • High-arched palate • Death: Respiratory failure • Foot drop • EOM spared	• Floppy infant/hypotonia • Ptosis • Extraocular muscle involvement • Facial diplegia • Dysphagia • Respiratory insufficiency	• Floppy infant/hypotonia • Hip contractures • Hip dislocations
Labs	M Bx: Central cores in Type I fibers Absent mitochondria	M Bx: Rod-shaped bodies on Gomori trichrome stain	M Bx: Central location of fiber nuclei, forming chains	M Bx: Numerous small Type I and normal to large Type II fibers
EDX findings	EMG • ER, SDSA MUAP	EMG • ER, SDSA MUAP	EMG • AA, ER, SDSA MUAP	EMG • ER, SDSA MUAP
Treatment	Bracing	Rehabilitation, surgery	Rehabilitation, antiseizure medication	Rehabilitation, bracing

AA, abnormal activity; EDX, electrodiagnostic; EMG, electromyography; EOM, extraocular muscles; ER, early recruitment; M Bx, muscle biopsy; MUAP, motor unit action potential; SDSA, short-duration, small-amplitude.

TABLE 5–52 Inflammatory Myopathies

DISORDER	POLYMYOSITIS/DERMATOMYOSITIS	INCLUSION BODY MYOSITIS
Etiology	Autoimmune, connective tissue disorder, infection, cancer	Unknown
Clinical presentation	• Symmetrical proximal weakness: Hips followed by shoulders • Neck flexion weakness • Myalgias, dysphagia, dysphonia • No facial or ocular muscle weakness • Dermatomyositis: Periorbital violet rash and edema Gottron's sign: Red-purple patches over the knuckles, elbows, knees	• Asymmetric, slowly progressive, painless weakness in proximal and distal muscles • Associated with a polyneuropathy • Affects adults 45–55 years and peaks at 70 years
Labs	Blood: Increased CPK, ESR, aldolase, SGOT, SGPT, LDH M Bx: Necrosis of the Type I and II fibers. Perifascicular atrophy.	Blood: Increase in CK M Bx: Rimmed or cytoplasmic/basophilic vacuoles. Eosinophilic inclusion bodies.
EDX findings	NCS • SNAP: Normal • CMAP: Normal EMG • 🕮 AA (most commonly in the paraspinals), ER, SDSA MUAP	NCS • SNAP: ± Abnormal • CMAP: ± Abnormal EMG • AA, ER, ± SDSA MUAP
Treatment	Rehabilitation. Medications: Corticosteroids, cytotoxic agents, IV immunoglobulin, plasmapheresis. Rest. Hydroxychloroquine for skin manifestations (dermatomyositis).	Rehabilitation. This condition is refractory to steroid treatment. No treatment.

AA, abnormal activity; CK, creatine kinase; CMAP, compound muscle action potential; CPK, creatine phosphokinase; EDX, electrodiagnostic; EMG, electromyography; ER, early recruitment; ESR, erythrocyte sedimentation rate; LDH, lactate dehydrogenase; M Bx, muscle biopsy; MUAP, motor unit action potential; NCS, nerve conduction study; SDSA, short-duration, small-amplitude; SGOT, serum glutamic oxaloacetic transaminase; SGPT, serum glutamic pyruvic transaminase; SNAP, sensory nerve action potential.

TABLE 5–53 Metabolic Myopathies: Common Disorders

CHARACTERISTICS	MCARDLE'S DISEASE (TYPE V)	POMPE'S DISEASE (TYPE II)
Etiology	Autosomal recessive *Myophosphorylase deficiency*	Autosomal recessive *Acid maltase deficiency*
Onset	<15 years of age	Infant to adult
Clinical presentation	• Exercise intolerance • Easy fatigability • Muscle stiffness • Cramping • Second-wind phenomenon: Brief rest improves symptoms • Strenuous exercise can precipitate myolysis (possibly cause renal failure and death)	• Hypotonia • Tongue enlargement • Cardiomegaly • Hepatomegaly • Respiratory insufficiency • Death by 2 years of age • A milder form may affect adults
Labs	Urine: Myoglobinuria M Bx: Excess glycogen, absent phosphorylase	Blood: Increase CK during the attacks M Bx: Vacuoles in Type I and II fibers
EDX findings	NCS • SNAP: Normal • CMAP: Normal EMG • Electrical silence during attacks (contracture)	NCS • SNAP: Normal • CMAP: Normal EMG • AA, ER, SDSA MUAP
Treatment	Supportive	Supportive

AA, abnormal activity; CK, creatine kinase; CMAP, compound muscle action potential; EDX, electrodiagnostic; EMG, electromyography; ER, early recruitment; M Bx, muscle biopsy; MUAP, motor unit action potential; NCS, nerve conduction study; SDSA, short-duration, small-amplitude; SNAP, sensory nerve action potential.

TABLE 5–54 Metabolic Myopathies Periodic Paralysis: Common Disorders

CHARACTERISTICS	HYPERKALEMIC PERIODIC PARALYSIS	HYPOKALEMIC PERIODIC PARALYSIS
Etiology	Autosomal dominant *Multiple secondary causes*	Autosomal dominant *Multiple secondary* causes
Onset	Childhood–second decade	Starts in early second decade
Clinical presentation	• Proximal muscle weakness • Paresthesias of the lips and lower limbs • Myotonia • Attacks last 10–60 minutes • May be aborted with exercise • Exacerbated with cold exposure and rest following exercise	• Weakness starts in the legs and spreads proximally • Attacks last 12–24 hours • Myotonia seen in the eyelids • Exacerbated with rest after exercise, stress, and a high carbohydrate diet
Labs	Blood: High K+ during the attack	Blood: Low potassium M Bx: Normal
EDX findings	NCS • SNAP: Normal • CMAP: Normal EMG • During an attack: ER, SDSA MUAP, AA	NCS • SNAP: Normal • CMAP: Normal EMG • During an attack: Electrical silence
Treatment	Diet: High carbohydrate	Diet: K+ supplement

AA, abnormal activity; CMAP, compound muscle action potential; EDX, electrodiagnostic; EMG, electromyography; ER, early recruitment; M Bx, muscle biopsy; MUAP, motor unit action potential; NCS, nerve conduction study; SDSA, short-duration, small-amplitude; SNAP, sensory nerve action potential.

TABLE 5–55 Myopathies: Common Disorders

CHARACTERISTICS	MYOTONIA CONGENITA (THOMSEN'S DISEASE; LITTLE HERCULES)	PARAMYOTONIA CONGENITA (EULENBURG)
Etiology	Autosomal dominant	Autosomal dominant
Onset	Birth–adulthood	Birth–adulthood
Clinical presentation	• Severe spasms exacerbated by the cold • Improves with warmth and exercise • Muscle hypertrophy • Myotonia • No weakness	• Stiffness • Weakness • Fatigue • Myotonia • Exacerbated with cold and exercise
Labs	Blood: CK—normal	M Bx: Fiber size variation
EDX findings	NCS • SNAP: Normal • CMAP: Normal EMG • AA (myotonic discharges, no fibs, no pos. waves), normal recruitment, normal MUAP	NCS • SNAP: Normal • CMAP: Decreased with cooling EMG • Electric silence or AA with cooling.
Treatment	Medication: Procainamide, Dilantin, quinine (PDQ)	Warm extremities

AA, abnormal activity; CK, creatine kinase; CMAP, compound motor action potential; EDX, electrodiagnostic; EMG, electromyography; M Bx, muscle biopsy; MUAP, motor unit action potential; NCS, nerve conduction study; SNAP, sensory nerve action potential.

TABLE 5–56 Steroid Myopathy

CHARACTERISTICS	STEROID MYOPATHY	STATIN MYOPATHY
Etiology	Corticosteroid proteolysis effect	Multifactorial
Onset	Weeks to years post use	Weeks to years post use
Clinical presentation	• Proximal muscle weakness • Risk is increased if on 30 mg/day • Preferentially affects the hip girdle muscles	• Proximal weakness and pain • Worse with exercise • Lipophilic statins (simvastatin, atorvastatin, lovastatin) are more likely to produce muscular effects than are relatively hydrophilic agents (pravastatin, rosuvastatin, fluvastatin)
Labs	M Bx: Type II atrophy	M Bx: Nonspecific
EDX findings	NCS • SNAP: Normal • CMAP: Normal EMG • May be abnormal with small polyphasics in very severe cases	NCS • SNAP: Normal • CMAP: Normal EMG • May be abnormal with small polyphasics in very severe cases
Treatment	Rehabilitation, exercise, stop steroids	Stop statins, or stop medications that interfere with cytochrome-P450: Macrolide antibiotics, verapamil, diltiazem, cimetidine, and so forth

CMAP, compound motor action potential; EDX, electrodiagnostic; EMG, electromyography; M Bx, muscle biopsy; NCS, nerve conduction study; SNAP, sensory nerve action potential.

MOTOR NEURON DISEASES

ETIOLOGY

- This is a disorder resulting from the progressive degeneration of the motor neurons in the spinal cord, brainstem, or motor cortex (Table 5–57).
- It manifests as muscular weakness and atrophy with varying corticospinal tract signs. See Chapter 10, Pediatric Rehabilitation for further information.

TABLE 5–57 Motor Neuron Diseases

LOWER MOTOR NEURON LESION	UPPER AND LOWER MOTOR NEURON LESION	UPPER MOTOR NEURON LESION
SMA Poliomyelitis/Post-polio syndrome	ALS	PLS Hereditary spastic paraplegia

ALS, amyotrophic lateral sclerosis; PLS, primary lateral sclerosis; SMA, spinal muscle atrophy.

CLINICAL PRESENTATION (TABLE 5–58)

TABLE 5–58 Lower Versus Upper Motor Neuron Signs

LOWER MOTOR NEURON SIGNS	UPPER MOTOR NEURON SIGNS
Atrophy Flaccidity Hyporeflexia Fasciculations	Weakness Spasticity Hyperreflexia Upgoing plantar response

ELECTRODIAGNOSTIC FINDINGS

- Evaluation should include at least one upper and one lower limb, starting in the most severely affected muscles. It is important to rule out treatable neuropathies that mimic motor neuron disease (MND). An example of this is multifocal motor neuropathy, which includes conduction block and temporal dispersion, not seen in MND.

NCS

- SNAP: Typically normal. Abnormal can be seen in hereditary spastic paraplegia and spinobulbar muscular atrophy.
- CMAP: Variable findings
  - Abnormalities in weakened muscles with asymmetric side-to-side comparisons demonstrating decreased CV and prolonged latencies.
- F-wave: Abnormal latency, persistence with chronodispersion

EMG

- Classic findings are seen in the area of clinical UMN and lower motor neuron (LMN) abnormalities. Active denervation with reinnervation must be found in three of four body segments (craniobulbar, cervical, thoracic, and lumbosacral). At least two muscles with different innervations should be abnormal.
- Chronic neurogenic changes (decreased MUAP recruitment, large-amplitude MUAP, rapidly firing potential) and denervation potentials (FIB, PSWs, fasciculations) should be evident (Table 5–59).
- Recruitment is decreased. There are polyphasic potentials, due to reinnervation.
- SFEMG: Abnormal FD, jitter, and blocking.

TABLE 5–59 Abnormal Spontaneous Activity in Motor Neuron Disease

FIBS AND PSWS	FASCICULATIONS	CRDS
• SMA Type I • SMA Type II • SMA Type III • ALS • Poliomyelitis	• ALS • Poliomyelitis • Post-polio syndrome	• SMA Type III

ALS, amyotrophic lateral sclerosis; CRD, complex repetitive discharge; FIB, fibrillation potential; PSW, positive sharp waves; SMA, spinal muscular atrophy.

Recruitment of the Diaphragm

A monopolar needle recording electrode is inserted through the eighth or ninth intercostal space at the anterior axillary line. MUAP of the intercostal muscles is recruited during expiration and is higher in amplitude than the diaphragm, which is recruited during inspiration.

MUAP, motor unit action potential.

The following tables outline pertinent MND patterns. Please refer to Table 5–57 as an overview for Tables 5–60 and 5–61.

TABLE 5–60 Motor Neuron Diseases: Spinal Muscular Atrophy Types I, II, and III

CHARACTERISTICS	SPINAL MUSCULAR ATROPHY TYPE I (WERDNIG-HOFFMAN DISEASE)	SPINAL MUSCULAR ATROPHY TYPE II	SPINAL MUSCULAR ATROPHY TYPE III (KUGELBERG–WELANDER DISEASE)
Genetic etiology	Autosomal recessive	Autosomal recessive	Autosomal recessive/ dominant
Onset	3–6 months	2–12 months	2–15 years
Course	Death by 2–3 years of age	Death by approximately 10 years old	Normal life expectancy
	Worst prognosis	Wheelchair by 2–3 years of age	Wheelchair by 30 years of age
Progression	Rapid; fatal (respiratory failure)	Slower; fatal (respiratory failure)	Slowly
Clinical presentation	• Floppy baby/hypotonia • Unable to reach milestones • Progressive weakness • Absent MSR • Difficulty feeding • Weak cry • Frog-legged position • Tongue fasciculations • 🕮 Facial muscle affected least • Extraocular muscles intact • Sphincter muscles are spared • Paradoxical breathing • 🕮 Never sits independently	• Floppy baby/hypotonia • Gradual progressive limb weakness; upper > lower • Absent MSR • Face least affected • Kyphoscoliosis • Equinus deformity of the feet • ± Tongue fasciculations • Progressive pulmonary involvement • 🕮 Independent sitting • 🕮 Assistive devices for standing and walking	• Symmetric weakness: Lower limb then upper limb • Abnormal MSR • 🕮 ± Gowers' sign • 🕮 ± Calf pseudohypertrophy • ± Dysphagia • ± Dysarthria • Tongue fasciculations—late onset • Normal intelligence • 🕮 Independent standing/ walking

(*continued*)

TABLE 5–60 Motor Neuron Diseases: Spinal Muscular Atrophy Types I, II, and III (*continued*)

CHARACTERISTICS	SPINAL MUSCULAR ATROPHY TYPE I (WERDNIG-HOFFMAN DISEASE)	SPINAL MUSCULAR ATROPHY TYPE II	SPINAL MUSCULAR ATROPHY TYPE III (KUGELBERG–WELANDER DISEASE)
Labs	Blood: Increase CPK levels M Bx: Hyper/atrophic fibers	Blood: Increase CPK levels M Bx: Hyper/atrophic fibers	Blood: Increase CPK levels M Bx: Hyper/atrophic fibers
EDX findings	NCS • SNAP: Normal • CMAP: ± Abnormal EMG • AA, LDLA/SDSA MUAP, DR	NCS • SNAP: Normal • CMAP: ± Abnormal EMG • AA, SDSA MUAP, DR	NCS • SNAP: Normal • CMAP: Normal EMG • AA, LDLA/SDSA MUAP, DR
Treatment	Supportive	Supportive rehabilitation	Supportive rehabilitation

AA, abnormal activity; CMAP, compound muscle action potential; CPK, creatine phosphokinase; DR, delayed recruitment; EDX, electrodiagnostic; EMG, electromyography; LDLA, long-duration, large-amplitude; M Bx, muscle biopsy; MSR, muscle stretch response; MUAP, motor unit action potential; NCS, nerve conduction study; SDSA, short-duration, small-amplitude; SNAP, sensory nerve action potential.

TABLE 5–61 Motor Neuron Disease: ALS, Polio, Post-Poliomyelitis

CHARACTERISTICS	ALS	POLIOMYELITIS	POST-POLIOMYELITIS SYNDROME
Pathology	Degeneration of the anterior horn cells	Degeneration of anterior horn cells	Loss of the anterior horn cells
Etiology	Unknown	Picornavirus orally enters the body and spreads via lymphoid system leading to orphaned muscle fibers	Death of the motor neuron due to aging Burnout of motor unit from increased metabolic demand (see Figure 5–128)
Clinical presentation	• Most commonly in men after the sixth decade • 🕮 First signs: Asymmetric atrophy, weakness, fasciculations • 🕮 Dysphagia (oral, pharyngeal), dysarthria • Pseudobulbar signs: A cluster of symptoms including difficulty chewing, swallowing, and speech, along with unprovoked emotional outbursts (e.g., crying and laughing) • 🕮 Bowel and bladder are spared • Sensation is spared • Extraocular muscles are spared • Upper and lower motor neuron signs • Prognosis: 50% die within 3 years, 30% live for 5 years, 10% live for 10 years	• Signs of infection: Fever, malaise, sore throat, vomiting, headache, back and neck pain, and stiffness • Weakness • Absent MSR • Bulbar palsies: Dysphasia, nasal voice • Sensation is spared • Autonomic dysfunction can occur • Prognosis: Disease can progress or remit – 25%: Severe disability – 25%: Mild disability – 50%: Complete recovery • Mortality: 1%–4% chance in children; 10% chance in adults with bulbar and respiratory involvement	Halstead-Ross Criteria 1. History of a previous diagnosis 2. Recovery of function 3. Stability for approximately 15 years 4. Return of symptoms 5. No other medical problems to explain new symptoms: – Weakness – Atrophy 6. Difficulties with ADLs – Fatigue – Arthralgia – Myalgia – Cold intolerance

(*continued*)

TABLE 5–61 Motor Neuron Disease: ALS, Polio, Post-Poliomyelitis (*continued*)

CHARACTERISTICS	ALS	POLIOMYELITIS	POST-POLIOMYELITIS SYNDROME
Clinical presentation *(cont.)*	• Wheelchair by 12–18 months • Predictors of survival: – Age of onset (younger is better) – Severity of onset – Pulmonary function – Abnormal sniff test correlates with poor survival		
EDX findings	NCS • SNAP: Normal • CMAP: Normal EMG • AA, DR, LDLA MUAP, CRDs LRRS • Increased decrement SFEMG • Increased jitter and fiber density Protocol: Abnormal activity in two muscles from two different nerve roots in three different body regions Body regions: Brainstem, cervical, thoracic, lumbar	NCS • SNAP: Normal • CMAP: Normal or decreased EMG • AA, DR, LDLA MUAP	NCS • SNAP: Normal • CMAP: Abnormal EMG • AA, DR, GIANT MUAP LRRS • Normal activity SFEMG • Increased jitter, fiber density, and blocking Post-poliomyelitis syndrome electrophysiologically resembles old stable poliomyelitis. Its diagnosis is not based on EMG/NCS but on clinical presentation.
Treatment	Rehabilitation, prevent contractures, submaximal exercise, tracheostomy, respiratory therapy, riluzole (Rilutek®) antiglutamate slows disease progression, prolongs ventilator time, BiPAP	Rehabilitation, pain management, prevent contractures	Rehabilitation, assistive devices, energy conservation, psychological counseling, avoid fatigue

AA, abnormal activity; ADLs, activities of daily living; ALS, amyotrophic lateral sclerosis; BiPAP, bilevel positive airway pressure; CMAP, compound muscle action potential; CRD, complex repetitive discharge; DR, delayed recruitment; EDX, electrodiagnostic; EMG, electromyography; LDLA, long-duration, large-amplitude; LRRS, low-rate repetitive stimulation; MSR, muscle stretch response; MUAP, motor unit action potential; NCS, nerve conduction studies; SNAP, sensory nerve action potential; SFEMG, single-fiber electromyography.

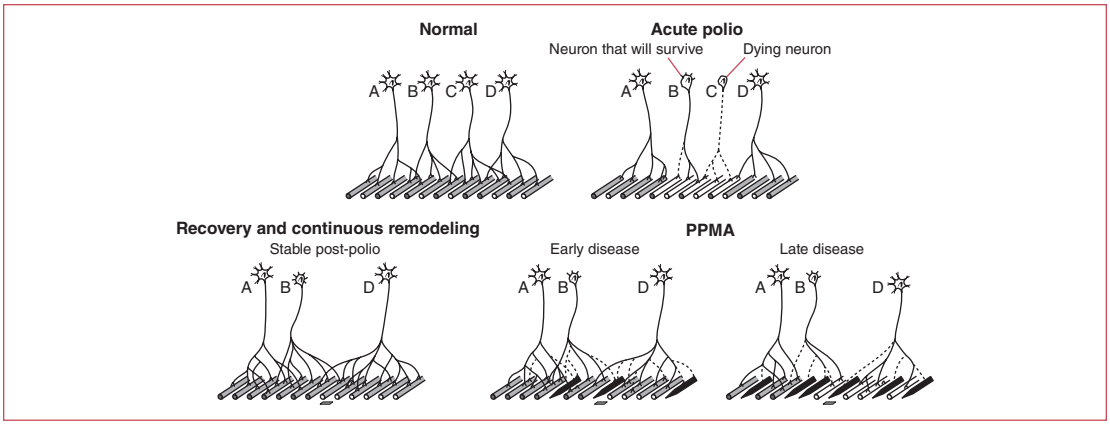
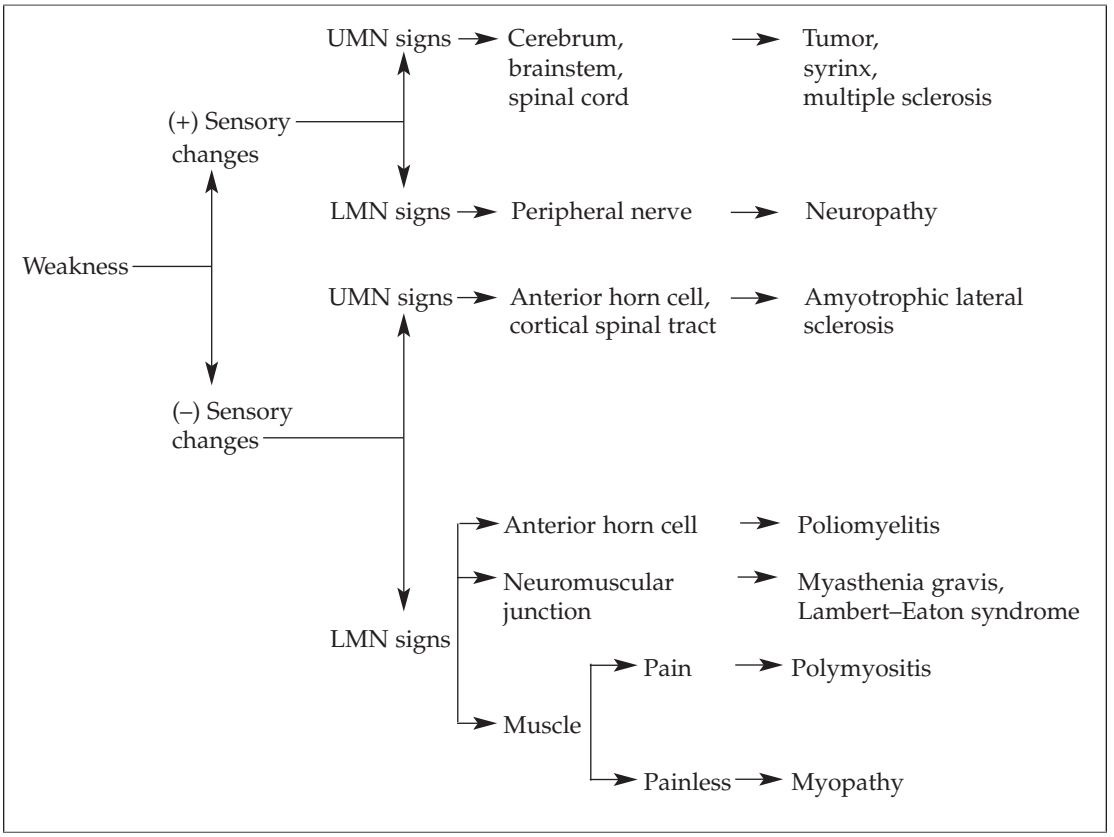


FIGURE 5–128 Post-poliomyelitis progressive muscular dystrophy. PPMA, progressive post-polio muscular atrophy.

WEAKNESS: DIFFERENTIAL DIAGNOSIS

TABLE 5–62 Differential Diagnosis of Weakness



LMN, lower motor neuron; UMN, upper motor neuron.

CRITICAL ILLNESS NEUROMUSCULAR DISEASE (TABLE 5–63)

- In the ICU patient, various factors including infection, trauma, surgery, chemical exposure, and sepsis can lead to systemic inflammatory response syndrome (SIRS). Single or multiple organ failure can ensue. Septic encephalopathy and critical illness polyneuropathy (CIP) may result. Neuromuscular blocking agents and steroids added to this may cause critical illness myopathy (CIM).
- SIRS is present if two or more of the following occurs:
  - Body temperature >38°C or <36°C
  - Heart rate >90 beats per minute (bpm)
  - Tachypnea:
    - Respiratory rate >20 breaths per minute
    - Hypocapnia $PaCO_2$ <32 torr (<4.3 KPa)
  - White blood cell (WBC) >12,000 or <4,000, or >10% immature neutrophils (bands)
- CIP is the degradation of neural tissue due to multiple medical complications causing a primarily axonal as well as demyelinating motor and sensory peripheral polyneuropathy.
- CIM is an acute, generally inflammatory myopathy due to multiple medical complications causing muscle membrane instability and muscle cell breakdown.
- Bilateral diffuse weakness predominant in the proximal part of the limbs after improvement of the acute phase of the critical illness is highly suggestive of critical illness neuromuscular disease (Table 5–63).

TABLE 5–63 Critical Illness Neuromuscular Disease

DISORDER	SEPTIC ENCEPHALOPATHY	CRITICAL ILLNESS POLYNEUROPATHY	CRITICAL ILLNESS MYOPATHY
Etiology	SIRS	SIRS	SIRS
Clinical presentation	Altered mental status, an early complication of SIRS	Difficulty weaning from the ventilator; flaccid limbs	Respiratory weakness; flaccid limbs
Labs	Near-normal CSF; may have normal head scans, suggesting functional vs. structural impairment	Normal CK; muscle biopsy; denervation atrophy; other labs altered as with SIRS	Variably elevated; muscle biopsy; loss of thick filaments; other labs altered as with SIRS
EDX		Motor and sensory NCS: Axonal loss pattern or absent EMG: Distal pattern of decreased recruitment with or without denervation and reinnervation, depending on the time course	Motor NCS: Low amplitudes throughout Sensory NCS: Normal EMG: Small, short, polyphasic MUAPs with normal or early recruitment; active denervation may be present

CK, creatine kinase; CSF, cerebrospinal fluid; EDX, electrodiagnostic; EMG, electromyography; MUAP, motor unit action potential; NCS, nerve conduction study; SIRS, systemic inflammatory response syndrome.

Treatment

- Multifactorial and difficult. Treat underlying sepsis and multiorgan failure. Supportive care.
- Other disorders of muscle in the differential in the ICU patient: Rhabdomyolysis necrotizing myopathy, disuse myopathy.
- Other disorders to differentiate from CIP: Spinal cord compression, MND, Guillain–Barré syndrome, MG, myasthenic syndrome.

CENTRAL NERVOUS SYSTEM DISORDERS

- In CNS disorders, sensory and motor NCS are normal.
- On needle EMG, there is no denervation or reinnervation on the weak limb(s); it reveals normal spontaneous activity and normal MUAP morphology. On voluntary contraction, however, the interference pattern is not complete due to decreased activation (i.e., decreased firing frequency); the number of available MUAPs (i.e., recruitment) remains normal.
- In a segmental spinal cord lesion, muscle innervated below the level of lesion will display the typical pattern of CNS lesion (i.e., decreased activation). At the segmental level of the lesion, anterior horn cells may be affected, leading to a neuropathic pattern in muscles innervated by that segment.

PARANEOPLASTIC NEUROLOGIC SYNDROMES

- Paraneoplastic neurologic syndromes (PNS) result from immune cross-reactivity between tumor cells and components of the nervous system.
- Paraneoplastic Syndromes are detected before cancer is diagnosed in 80% of cases.
- Although Paraneoplastic Syndromes are rare, affecting <1% of cancer patients overall, certain malignancies have a substantially higher incidence of these conditions. Up to 5% of patients with small cell lung cancer and up to 10% of patients with lymphoma or myeloma develop Paraneoplastic Syndrome.
- Paraneoplastic Syndromes can affect the CNS (e.g., limbic encephalitis and paraneoplastic cerebellar degeneration), the NMJ (e.g., LEMS and MG), or the peripheral nervous system (e.g., autonomic neuropathy and subacute sensory neuropathy).

- The diagnosis of PNS may incorporate imaging, serologies, EEG, NCS, EMG, and cerebrospinal fluid analysis.
- Because most patients diagnosed as having an apparent PNS will not have known cancer at the time, screening for an underlying tumor is indicated.
- Immunosuppressive therapy is a mainstay of PNS treatment, but success is variable.

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PROSTHETICS AND ORTHOTICS

6

Heikki Uustal, MD • Edgardo Baerga, MD • Jaclyn Joki, MD •
Leslie Bagay, MD • Yi (Dan) Zhou, MD

GAIT ANALYSIS

TERMINOLOGY

The Gait Cycle (Figures 6–1 and 6–2; Tables 6–1 to 6–3)

- A single sequence of functions of one limb is called a gait cycle. It is essentially the functional unit of gait.
- One gait cycle is also referred to as a stride. Each stride comprises two steps:
  1. **Stride length:** Linear distance between corresponding successive points of contact of the same foot. It is the distance measured from heel strike to heel strike of the same foot (Figure 6–1).
  2. **Step length:** Linear distance in the plane of progression between corresponding successive contact points of opposite feet. It is the distance measured from heel strike of one foot to heel strike of the other foot. Normally, the step length is approximately 15 to 20 in. (15 in. = 38 cm; Figure 6–1).

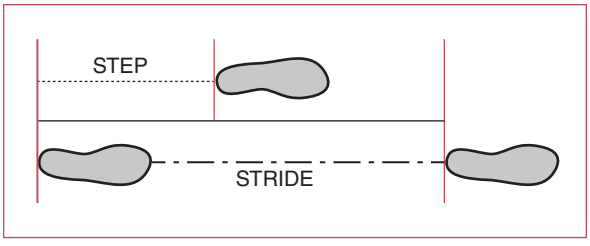


FIGURE 6–1 Step and stride.

- The gait cycle comprises two phases, stance phase and swing phase:
  - **Stance phase:** Time period during which the limb is in contact with the ground. Stance phase is divided into five periods (see Stance Phase: Five Subdivisions).
  - **Swing phase:** Time period during which the foot is in the air for limb advancement. Swing phase is divided into three periods (see Swing Phase: Three Subdivisions).
- The normal distribution of time during the gait cycle at normal walking speed is 60% for *stance phase* and 40% for *swing phase*. Walking faster decreases the time spent in stance phase and increases time spent in swing phase (Figure 6–2C).
- ***Double-limb support:*** Time period during which both feet are in contact with the floor. Both the beginning and end of the stance phase are considered to be the double-limb support period.
  - Usually comprises 20% of the normal gait cycle versus 80% of single-limb support
- ***Single-limb support:*** Time period starts when the opposite foot is lifted for the swing phase
  - The amount of time spent during double-limb support decreases as the speed of walking increases. When there is no longer a double-limb support period, the person is considered to be running.
- **Cadence:** Number of steps per unit of time

- Comfortable walking speed = 80 m/min or 3 mph. Speed slows by either reducing the cadence or by decreasing step or stride length.
- 🕮 **Center of gravity (COG):** Typically located 5 cm anterior to the S2 vertebra. The COG is displaced 5 cm (<2 in.) horizontally and 5 cm vertically during an average adult male step.
- **Base of support:** Defined as the space outlined by the feet and any assistive device in contact with the ground. Falling is avoided if the COG remains positioned over the base of support.
  - Normal base of support (distance between heels) = 6 to 10 cm

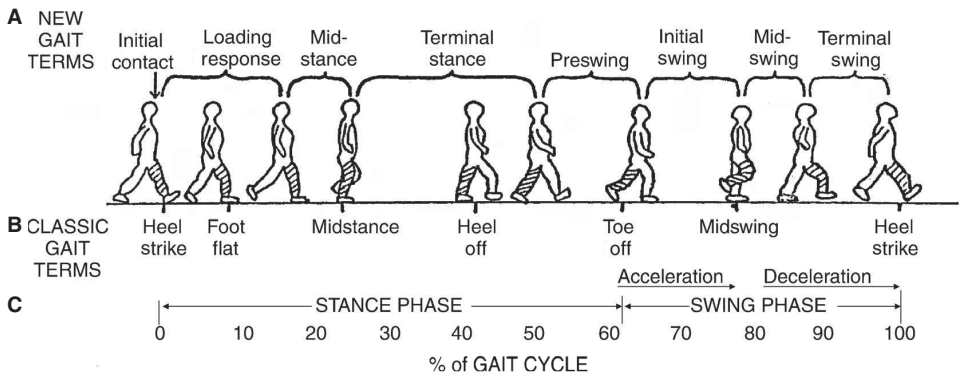


FIGURE 6–2 Gait cycle: (A) New gait terms. (B) Classic gait terms. (C) The normal distribution of time during the gait cycle at normal walking speed.
Source: Courtesy of Carson Schneck, MD.

STANCE PHASE: FIVE SUBDIVISIONS (TABLE 6–1; FIGURE 6–2)

- Remember: **"I Like My Tea Presweetened."**
  1. **Initial contact:** Instant the foot contacts the ground
  2. **Loading response:** Time period immediately following initial contact up until the contralateral extremity is lifted off the ground, during which weight shift occurs
     🕮 The body has the lowest COG during loading response.
  3. **Midstance:** Time period from lift of the contralateral extremity from the ground to the point where the ankles of both extremities are aligned in the frontal (or coronal) plane
     🕮 The body has the highest COG during midstance.
  4. **Terminal stance:** Time period from ankle alignment in the frontal plane to just prior to initial contact of the contralateral (swinging) extremity
  5. **Preswing:** Time period from initial contact of the contralateral extremity to just prior to lift of the ipsilateral extremity from the ground (unloading weight)

🕮 **TABLE 6–1 Gait Cycle Phases/Events**

NEW TERMINOLOGY (FIGURE 6–2A)	TRADITIONAL TERMINOLOGY (FIGURE 6–2B)	
Initial contact	Heel (foot) strike	STANCE PHASE
Loading response	Foot flat	
Midstance	Midstance	
Terminal stance	Heel off	
Preswing	Toe off	
Initial swing	Acceleration	SWING PHASE
Midswing	Midswing	
Terminal swing	Deceleration	

SWING PHASE: THREE SUBDIVISIONS (TABLE 6–1; FIGURE 6–2)

- Remember: "**I**n **M**y **T**eapot"
  1. **Initial swing:** Lift of the extremity from the ground to position of maximum knee flexion
  2. **Midswing:** Immediately following knee flexion to vertical tibia position
  3. **Terminal swing:** Following vertical tibia position to just prior to initial contact

DETERMINANTS OF GAIT (TABLE 6–2)

- These factors are applied in normal human gait to minimize the excursion of the body's COG and help produce forward progression with the least expenditure of energy (Saunders et al., 1953). The six determinants of gait are:
  1. Pelvic rotation
  2. Pelvic tilt
  3. Knee flexion in stance phase
  4. Foot mechanisms
  5. Knee mechanisms
  6. Lateral displacement of the pelvis

TABLE 6–2 Determinants of Gait

DETERMINANTS OF GAIT	
1. Pelvic rotation	The pelvis rotates medially (anteriorly) on the swinging leg side, lengthening the limb as it prepares to accept weight. With 4–5 degrees of pelvic rotation in either direction during double support, the limbs are essentially lengthened in the would-be lowest point of the gait cycle (preventing a sudden drop of the COG).
2. Pelvic tilt	The pelvis on the side of the swinging leg (opposite to the weight-bearing leg) is lowered 4–5 degrees. This lowers the COG at midstance.
3. Knee flexion in stance	Early knee flexion → knee flexion at foot strike (15 degrees). The bending of the knee reduces the vertical elevation of the body at midstance (would-be highest point in the gait cycle) by shortening the hip-to-ankle distance. This lowers the COG (by minimizing its vertical displacement), decreasing the energy expenditure. It also tends to absorb the shock of impact at heel strike by lengthening (eccentric) contraction of the quadriceps.
4. Foot mechanisms (ankle flexion/extension mechanisms)	At heel strike, ankle plantar flexion smoothens the curve of the falling pelvis. It is associated with controlled plantar flexion during the first part of the stance. Three pivot points (rockers) at the heel, ankle, and forefoot help to functionally lengthen the stance limb at initial contact and preswing, and shorten the limb at midstance.
5. Knee mechanisms	After midstance, the knee extends as the ankle plantar flexes and the foot supinates to restore the length to the leg and diminish the fall of the pelvis at the opposite heel strike.
6. Lateral displacement of the pelvis	There is displacement toward the stance limb. The net COG of the body must lie above the base of support (the stance foot); therefore, a narrow base of support will minimize lateral motion.

Note: Determinants 1 to 5 reduce displacement on the vertical plane. Determinant 6 reduces displacement on the horizontal plane.
COG, center of gravity.

GAIT PATHOLOGY AND PROBABLE CAUSES

Tables 6–3 and 6–4 delineate major muscle activity and potential causes of abnormalities during the gait cycle.

TABLE 6–3 Major Muscle Activity During Gait Cycle

THE MAJOR MUSCLE ACTIVITY DURING EACH PHASE OF THE GAIT CYCLE VARIES DRAMATICALLY WITH RELATION TO WHETHER THE MUSCLES ARE INACTIVE, CONCENTRICALLY CONTRACTING, OR ECCENTRICALLY CONTRACTING.								
CLASSIC GAIT TERMINOLOGY	HEEL STRIKE	FOOT FLAT	MIDSTANCE	HEEL OFF	TOE OFF	ACCELERATION	MIDSWING	DECELERATION
NEW TERMINOLOGY (RANCHO LOS AMIGOS)	INITIAL CONTACT	LOADING RESPONSE	MIDSTANCE	TERMINAL STANCE	PRESWING	INITIAL SWING	MIDSWING	TERMINAL SWING
	STANCE PHASE 60%				SWING PHASE 40%			
% of total phase	0%–2%	0%–10%	10%–30%	30%–50%	50%–60%	60%–73%	73%–87%	87%–100%
Iliopsoas	Inactive	Inactive	Inactive	Concentric	Concentric	Concentric	Concentric	Inactive
Gluteus maximus	Eccentric	Inactive	Inactive	Inactive	Inactive	Inactive	Inactive	Inactive
Gluteus medius	Eccentric	Eccentric	Eccentric	Eccentric	Inactive	Inactive	Inactive	Inactive
Hamstrings	Eccentric	Eccentric	Inactive	Inactive	Inactive	Eccentric	Eccentric	Eccentric
Quadriceps	Eccentric	Eccentric[a]	Inactive	Inactive	Eccentric	Eccentric	Inactive	Inactive
Pretibial muscles	Eccentric	Eccentric	Inactive	Inactive	Inactive	Concentric	Concentric	Concentric
Calf muscles	Inactive	Inactive	Eccentric	Concentric	Concentric	Inactive	Inactive	Inactive

Key: Inactive Concentric Eccentric

[a]The quadriceps change from an eccentric to concentric contraction as it transitions from loading response to midstance.

TABLE 6–4 Gait Pathology and Probable Causes

GAIT PATHOLOGY	PROBABLE CAUSES
1. **Initial contact**	
– Foot slap	• Moderately weak ankle dorsiflexors (grade 3/5)
2. **Initial contact through midstance**	
– Genu recurvatum	• Weak, short, or spastic quadriceps; compensated hamstring weakness; Achilles tendon contracture; plantar flexor spasticity
– Excessive foot supination	• Compensated forefoot valgus deformity; pes cavus; short limb
– Excessive trunk extension	• Weak hip extensor or flexor; hip pain; decreased knee ROM
– Excessive trunk flexion	• Weak gluteus maximus and quadriceps; hip flexion contracture
3. **Initial contact through preswing**	
– Excessive knee flexion	• Hamstring contracture; increased ankle dorsiflexion; weak plantar flexor; long limb; hip flexion contracture
– Excessive medial femur rotation	• Tight medial hamstrings; anteverted femoral shaft; weakness of opposite muscle group
– Excessive lateral femur rotation	• Tight lateral hamstrings; retroverted femoral shaft; weakness of opposite muscle group
– Wide base of support	• Hip abductor muscle contracture; instability; genu valgum; leg length discrepancy
– Narrow base of support	• Hip adductor muscle contracture; genu varum
4. **Loading response through preswing**	
– 🕮 Excessive trunk lateral flexion (compensated Trendelenburg gait)	• Ipsilateral gluteus medius weakness; hip pain
– Pelvic drop (uncompensated Trendelenburg gait)	• Ipsilateral gluteus medius weakness
– Waddling gait	• Bilateral gluteus medius weakness
5. **Midstance through preswing**	
– Excessive foot pronation	• Compensated forefoot or hindfoot varus deformity; uncompensated forefoot valgus deformity; pes planus; decreased ankle dorsiflexion; increased tibial varum; long limb; uncompensated internal rotation of tibia or femur; weak tibialis posterior
– Bouncing or exaggerated	• Achilles tendon contracture; gastroc-soleus spasticity plantar flexion
– Insufficient push-off	• Gastroc-soleus weakness; Achilles tendon rupture; metatarsalgia; hallux rigidus
– Inadequate hip extension	• Hip flexor contracture; weak hip extensor
6. **Swing phase**	
– Steppage gait/foot drop	• Severely weak dorsiflexors; equinus deformity; plantar flexor spasticity
– Circumduction	• Long limb; abductor muscle shortening or overuse, stiff knee
– Hip hiking	• Long limb; weak hamstring; quadratus lumborum shortening, stiff knee

ROM, range of motion.
Source: Adapted from Tan JC. *Practical Manual of Physical Medicine and Rehabilitation: Diagnostics, Therapeutics, and Basic Problems*. Mosby; 1998:65.

Trendelenburg Gait (Gluteus Medius Gait)

- When the hip abductor muscles (gluteus medius and minimus) are weak, the pelvic/trunk stabilizing effect of these muscles during gait is lost.
  - Example: Stand on right leg only—if pelvis drops on the left (contralateral) side (excessive pelvic tilt), then it is called an **uncompensated right Trendelenburg gait**. The contralateral pelvis drops because the ipsilateral hip abductors do not stabilize the pelvis to prevent the drop.
  - When a patient walks, if they lean their trunk to the right to prevent left (contralateral) pelvic drop, it is a **compensated Trendelenburg gait**. The patient exhibits an excessive lateral lean in which the trunk is thrust toward the weak side to keep the COG over the stance leg.
- In contrast to most other gait deviations, a Trendelenburg gait becomes less evident with faster walking due to a shorter stance phase (DeLisa et al., 2005).

🕮 Gait Impairment in Parkinson's Disease

- Gait disturbances in Parkinson's include stooped posture and festinating gait.
- Also characterized by shuffling, decreased arm swing, reduced trunk rotation, "start hesitation," and in severe instances freezing while walking or turning ("freezing gait")
- Reduced step length is the primary disturbance.

🕮 Gait Impairment in Duchenne Muscular Dystrophy

- Often characterized by toe walking:
  - Stance phase: Plantar flexion to maintain a weight line posterior to the hip and anterior to the extended knee
  - Compensatory for weak knee extensors, or for increased lumbar lordosis, which itself is compensating for weak hip extensors
- Waddling gait and trunk lean over weight-bearing limb may occur due to hip abductor weakness.

Gait Impairment in Plantar Flexion Weakness

- Can be seen in injuries such as Achilles tendon rupture, tibial nerve injury, S1 radiculopathy, poliomyelitis, and Charcot–Marie–Tooth disease
- Gait dysfunction characterized by excessive ankle dorsiflexion and persistent knee flexion during stance phase
- Decreased stride length due to decreased or absent heel rise, which also results in decreased walking speed and increased energy expenditure
- Ipsilateral pelvic drop in preswing, causing a lower COG, then increased energy consumption since the pelvis rises again during the stance phase of the normal contralateral leg
- Decreased or absent heel rise and lack of propulsion (DeLisa et al., 2005)

🕮 Gait Impairment With Hip Flexion Contracture

- Hip flexion contracture can alter normal gait patterns due to the decreased hip extension range of motion (ROM). This can result in compensation with increased anterior pelvic tilt, decreased contralateral step length, and increased knee flexion.
- It can also increase energy consumption: a 35-degree contracture due to iliopsoas tightness results in a 60% increase in energy consumption (DeLisa et al., 2005).

Energy Expenditure During Ambulation

ENERGY EXPENDITURE OF DIFFERENT LEVELS OF AMPUTATION/ETIOLOGIES VERSUS NORMAL CONTROLS DURING AMBULATION (TABLE 6–5)

- Energy demands required to return to normal ambulatory functions are high.
- Even the healthiest amputees cannot achieve normal gait in terms of velocity, cadence, or energy consumption.

TABLE 6–5 Energy Expenditure of Different Levels of Amputation Versus Normal Controls During Ambulation

LEVEL OF AMPUTATION	INCREASED METABOLIC COST ABOVE NORMAL
Syme's	15%
Traumatic TT BKA	25% (Short BKA—40%; Long BKA—10%)
Traumatic bilateral long BKA	41% (Gonzalez et al., 1974)
Traumatic TF AKA	60%–70%
Traumatic bilateral AKA	>200% (260%; Huang et al., 1979)
Traumatic AKA and BKA	118% net cost (Corcoran, 1971)
Vascular TT BKA	40%
Vascular TF AKA	100%
Hip disarticulation	100%–200%

AKA, above-the-knee amputation; BKA, below-the-knee amputation; TF, transfemoral; TT, transtibial.
Sources: From Corcoran PJ. Energy expenditure during ambulation. In: Downey JAR, Darling RC, eds. *Physiological Basis of Rehabilitation Medicine*. W. B. Saunders; 1971; Gonzalez EG, Corcoran PJ, Reyes RL. Energy expenditure in below-knee amputees: correlation with stump length. *Arch Phys Med Rehabil*. 1974;55:111–119; Huang CT, Jackson JR, Moore NB, et al. Amputation: energy cost of ambulation. *Arch Phys Med Rehabil*. 1979;60(1):18–24; Tan JC. *Practical Manual of Physical Medicine and Rehabilitation: Diagnostics, Therapeutics, and Basic Problems*. Mosby; 1998:65; Traugh GH, Corcoran PJ, Reyes RL. Energy expenditure of ambulation in patients with above-knee amputations. *Arch Phys Med Rehabil*. 1975;56:67–71.

WHEELCHAIR PROPULSION (Cerny et al., 1980)

- Measured the energy expenditure of wheelchair propulsion in patients with paraplegia
- In the subjects studied, there was only 9% increase in energy expenditure (compared with ambulation in normal subjects).

CRUTCH WALKING

- Crutch walking requires more energy than walking with a prosthesis.
- Muscles that need strengthening in preparation for crutch walking:
  - Latissimus dorsi
  - Triceps
  - Pectoralis major
  - Quads
  - Hip extensors
  - Hip abductors

AMPUTATION AND PROSTHETICS

DEFINITION

- A prosthesis is an artificial substitute for a missing body part.

EPIDEMIOLOGY

- In 1996, approximately 1.2 million persons were living with an amputation; prevalence was approximately 1.6 million in 2005, with projections that prevalence may double by the

year 2050 (Ziegler-Graham et al., 2008). A recent 2024 study however reported a prevalence estimated at 2.3 million in 2019, and a projected prevalence of over 5.6 million by 2060 (Rivera et al., 2024).

- It is estimated that 1 in 190 Americans are currently living with a loss of a limb (Ziegler-Graham et al., 2008).
- In 2019, there were over 560,000 amputations in the United States (Rivera et al., 2014).
- Studies estimate the 5-year mortality rate of all people with limb loss due to dysvascular disease to be between 50% and 74%.
- Patients with limb amputation have an increased risk of cardiovascular disease, obesity, joint and bone issues, and depression (Sheehan & Gondo, 2014; Varma et al., 2014).
- Main causes of amputation: Dysvascular disease (including diabetes and peripheral arterial disease [PAD]), trauma, cancer, and congenital issues.
  - Rates of cancer and trauma-related amputation are decreasing.
  - Rates of dysvascular amputation are *increasing*.
  - Incidence of congenital limb deficiencies is stable.
- Risk of limb loss increases with age (greatest risk age 65 or older).
- Amputation due to vascular disease:
  - Nearly half of the individuals who had an amputation due to vascular disease will die within 5 years.
  - Approximately 64% of all amputations due to dysvascular disease occur in those age 65 or older.
  - 75% to 93% of lower extremity (LE) amputations are the result of vascular disease.
  - African Americans with peripheral vascular disease (PVD) undergo amputation at two to four times the rate of Caucasians (Sheehan & Gondo, 2014; Varma et al., 2014).
- 🕮 Diabetes is a major risk factor, contributing to two-thirds of all LE amputations.
  - Among those with diabetes who have an LE amputation, up to 55% will require amputation of the second leg within 2 to 3 years.
  - The prevalence of PVD is 20% higher in the diabetic population.
  - Recent research shows a 67% decline in the *rate* of lower limb amputation from diabetes-related complications since 1996.
  - It is estimated that as many as 60% of diabetes-related amputations could have been prevented (Sheehan & Gondo, 2014; Varma et al., 2014).
  - 🕮 Treatment of diabetic ulcers involve timely debridement of necrotic tissue, removal of surrounding callus, infection management, and reduction in weight-bearing forces. Arterial insufficiency should be assessed in all patients with diabetic ulcer using the ankle–brachial index (ABI; Everett & Mathioudakis, 2018; Sinwar, 2015).
- 🕮 Trauma is the leading cause of upper extremity amputations, accounting for 80% of amputations, with the majority limited to digital amputations.
  - About 30,000 traumatic amputations occur in the United States every year.
  - Four out of five traumatic amputation victims are male, and most of them are between the ages of 15 and 30.
  - Motor vehicle collisions are the most common cause (51%), followed by machinery accidents (19%; Sheehan & Gondo, 2014; Varma et al., 2014).
  - Upper limb amputations account for approximately two-thirds of all trauma-related amputations.
- Various scoring systems have been developed to characterize the "mangled extremity," such as the Mangled Extremity Severity Score (**Table 6–6**), the Mangled Extremity Syndrome Index, the Predictive Salvage Index, and the Limb Salvage Index. Components vary but all contain the presence of vascular injury and limb ischemia as key variables (Bosse et al., 2001; Shores & Lee, 2013).
- Males are at higher risk for trauma-related amputations than females.

TABLE 6–6 Mangled Extremity Severity Score

VARIABLES	SCORE
SKELETAL/SOFT-TISSUE INJURY	
Low energy (stab; simple fracture; pistol gunshot wound)	1
Medium energy (open or multiple fractures, dislocation)	2
High energy (high speed MVA or rifle gunshot wound)	3
Very high energy (high speed trauma + gross contamination)	4
LIMB ISCHEMIA	
Pulse reduced or absent but perfusion normal	1[a]
Pulseless; paresthesias, diminished capillary refill	2[a]
Cool, paralyzed, insensate, numb	3[a]
SHOCK	
Systolic BP always >90 mmHg	0
Hypotensive transiently	1
Persistent hypotension	2
AGE (YEARS)	
<30	0
30–50	1
>50	2

[a]Score doubled for ischemia >6 hours.
BP, blood pressure; MVA, motor vehicle accident.
Source: From Topal AE, Eren MN, Celik Y. Lower extremity arterial injuries over a six-year period: outcomes, risk factors, and management. *Vasc Health Risk Manag*. 2010;6(1):1103–1110. doi:10.2147/VHRM.S15316.

UPPER LIMB AMPUTATIONS

Acquired Upper Limb Amputations

MOST COMMON CAUSES

1. Trauma is the leading cause of acquired amputation in the upper extremity (approximately 80%), occurring primarily in men ages 15 to 45 years.
2. Cancer/tumors
3. Vascular complications of diseases

Standard Levels of Upper Limb Amputation (Figure 6–3)

1. Transphalangeal
2. Transmetacarpal
3. Transcarpal
4. Wrist disarticulation
5. Transradial (below-elbow) amputation
6. Elbow disarticulation

7. Transhumeral (above-elbow) amputation—6.5 cm or more proximal to the elbow joint
8. Shoulder disarticulation
9. Forequarter amputation

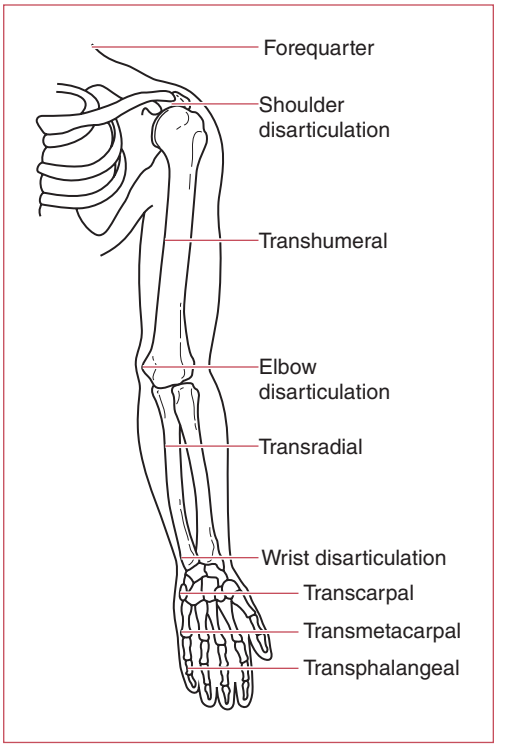


FIGURE 6–3 Upper limb levels of amputation (current terminology).

HAND/FINGER AMPUTATIONS (TRANSPHALANGEAL, TRANSMETACARPAL, TRANSCARPAL AMPUTATIONS)

- *Finger (transphalangeal) amputation* can occur at the distal interphalangeal (DIP), proximal interphalangeal (PIP), and metacarpophalangeal (MCP) levels.
- *Transmetacarpal amputation and wrist amputation* are seen less because they have decreased functional outcomes.
- *Multiple finger amputations*, including thumb and partial hand amputation, and those through the wrist, need to be considered carefully in view of the possible functional and cosmetic implications of prosthesis fitting and restoration. Inappropriate choice of amputation site can result in a prosthesis with disproportionate length or width.
- *Partial hand amputation* should be carefully planned to ensure adequate residual sensation and movement. For these amputations, a prosthesis may not be necessary. Surgical reconstruction may be a more appropriate choice of treatment to preserve or enhance function while maintaining sensation in the residual partial hand. There is little value in salvaging a partial hand with no prehension (ability to hold/grasp).
- 🕮 *Mangled hand:* Amputation is considered if irreparable damage occurs to four of the six basic parts (skin, vessels, skeleton, nerves, extensor, and flexor tendons). Initial goal: Save all feasible length.

WRIST DISARTICULATION

- A wrist disarticulation spares the distal radial ulnar articulation and thus preserves full forearm supination and pronation.
- Socket designs for this level are tapered and flattened distally to form an oval that allows the amputee full active supination and pronation, thus avoiding having to preposition the terminal device (TD) for functional activities.
- A special thin wrist unit is used to minimize the overall length of the prosthesis because of the extremely long residual limb.
- If cosmesis is of importance to the amputee, a long, below-elbow amputation may be a more appropriate amputation level.

TRANSRADIAL (BELOW-ELBOW) AMPUTATION (FIGURE 6–4)

- Transradial amputation is the most common level and allows a high level of functional recovery in the majority of cases.
- It can be performed at three levels:
  1. Very short: Residual limb length <35%
  2. Short: Residual limb length of 35% to 55%
  3. Long: Residual limb length of 55% to 90%

- The long below-elbow residual limb retains 60 to 120 degrees of supination and pronation, and the short below-the-elbow residual limb retains <60 degrees.
- The long forearm residual limb is preferred when optimal body-powered prosthetic restoration is the goal. It is the ideal level for the patient who is expected to perform physically demanding work.
- A residual limb length of 60% to 70% is preferred when optimal externally powered prosthetic restoration is the goal. This length typically permits good function and cosmesis while allowing adequate space for electronic components.
- The short and very short transradial amputation level can complicate suspension and limit elbow flexion strength and elbow ROM.

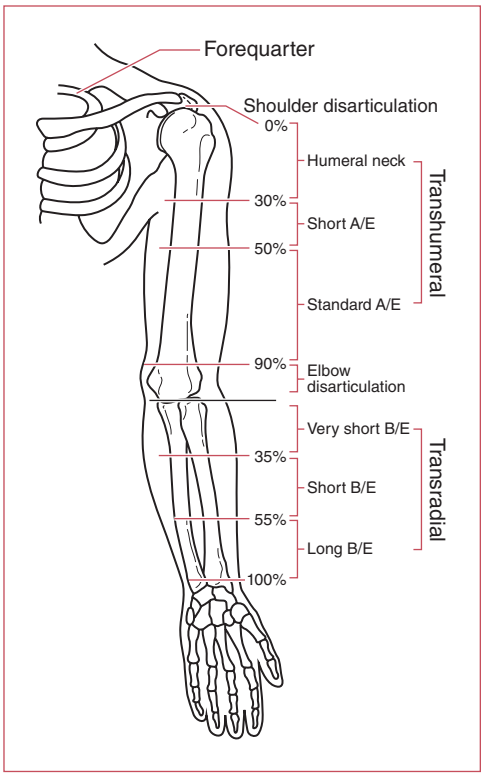


FIGURE 6–4 Levels of upper extremity amputation. A/E, above-elbow; B/E, below elbow.

ELBOW DISARTICULATION (FIGURE 6–4)

- Elbow disarticulation has surgical and prosthetic advantages and disadvantages.
- *Pros:* The surgical technique permits reduction in surgery time and blood loss, provides improved prosthetic self-suspension (by permitting the use of a less encumbering socket), and reduces the rotation of the socket on the residual limb compared with a transhumeral level of amputation. Allows greater lifting, pulling, and pushing power compared with above-elbow amputation.
- *Cons:* Major disadvantages are the marginal cosmetic appearance caused by the necessary external elbow mechanism, as well as the current limitations in technology that impede the use of externally powered elbow mechanisms at this level of amputation. These drawbacks often outweigh the advantages in the long run.
- In the patient for whom bilateral transhumeral amputation is the alternative, elbow disarticulation is the more desirable level when feasible in spite of the possible cosmetic problems.
- There is no risk of bone spur or heterotopic bone formation at the elbow disarticulation level.

TRANSHUMERAL (ABOVE-ELBOW) AMPUTATION (FIGURE 6–4)

- The transhumeral amputation can be performed at three levels:
  1. Humeral neck: Residual limb length of <30%
  2. Short transhumeral: Residual limb length of 30% to 50%
  3. Standard transhumeral: Residual limb length of 50% to 90%
- Longer residual limb length (up to 90% of humeral length) will give the best control and function with a prosthesis.
- These three amputation levels in most cases require similar prosthetic components, which can be externally powered, body-powered, passive, or have a combination of these.
- In above-elbow amputations with residual limb lengths >35%, usually the proximal trim line of the socket extends to within 1 cm of the acromion, and the socket is suspended by either a figure-8 or shoulder saddle and chest strap suspension system.
- Residual limbs shorter than 30% will be fitted as a shoulder disarticulation.

SHOULDER DISARTICULATION AND FOREQUARTER AMPUTATIONS (FIGURE 6–4)

- Fortunately, the shoulder disarticulation and forequarter amputations are seen with less frequency than amputations of other levels.

- In most cases, they are made necessary as part of the surgical intervention to remove a malignant lesion.
- Patients with these levels of amputation are the most difficult to fit with a functional prosthesis, due to the number of joints to be replaced and the problems related in maintaining secure suspension of the prosthesis.
- For shoulder disarticulation and forequarter amputations, the socket extends onto the thorax to suspend and stabilize the prosthesis.
- Prosthetic placement is more successful in those who are young, healthy, and male.
- Prosthetic components are similar to those of the transhumeral prosthesis, with the addition of the shoulder unit, which may be fixed, or passive positioning of the shoulder joint in flexion-extension and adduction-abduction.
- The joint may be provided with controls, in addition to the body-powered or externally powered control mechanisms needed for the elbow, wrist, and hand.
- Functional prosthetic use in the forequarter amputation is less successful because suspension is difficult to maintain. In some cases, a better option is to provide a passive, cosmetic prosthesis. Special consideration should be made for providing a shoulder cap to allow the patient to wear clothing more easily and improve cosmesis. The use of an ultralight passive prosthesis is usually well accepted in these patients.

UPPER LIMB PROSTHETICS

Terminal Devices and Wrist Units in Upper Extremity Prostheses

- Prosthetic components for upper extremity amputations include terminal devices, wrist units, sockets, elbow hinges, and shoulders.
- Terminal devices and wrist units are discussed in this section, while the rest of the componentry is discussed later in relation to specific amputation levels.

TERMINAL DEVICES

- Most patients who suffer an upper limb amputation and undergo prosthetic restoration require a terminal device for their prosthesis. They are used in all upper limb prostheses for amputations at the wrist level and above.
- Terminal devices lack sensory feedback and have limited mobility and dexterity.
- There are a variety of prosthetic terminal devices available and include passive, body-powered, and externally powered hooks and hands, and specialty devices for work or recreation.
  1. **Passive terminal devices:**
     - Lighter
     - Have no functional mechanisms and provide no grasp

     a. *Passive hand:*
        - Intended for cosmetic use only, but can also assist with pushing or lifting

     b. *Flexible passive terminal devices:*
        - Mitt-shaped terminal devices can fit into a glove or mitt for sports and other activities.

     c. *Specialty terminal devices* with adapters for sports equipment, hand tools, or kitchen utensils
  2. **Body-powered terminal devices** (hooks or hands) can be voluntary-opening (VO) or voluntary-closing (VC) types.
     - Prosthetic hands provide a three-jaw chuck pinch (three-jaw chuck involves grip with the thumb, index, and middle fingers).
     - Hooks provide the equivalent of lateral or tip pinch. In a normal hand, lateral or key grip involves contact of the pulp of the thumb with the lateral aspect of the corresponding finger.

     a. *VO terminal device:*
        - 🕮 Most common and practical type
        - Device maintained in closed position by rubber bands or tension springs
        - The amputee uses cable-control harness powered by proximal muscles to open the terminal devices against the force of the rubber bands or spring.

- To grasp, the patient releases the opened terminal device on an object; the rubber bands or spring provide the prehensile force.
- The pinch force is determined by the number and type of rubber bands or springs. Each rubber band provides about 1 lb. of pinch force.
- To control the amount of prehensile force, the patient must generate a continued opening force.

b. *VC terminal device:*
- More physiologic function than VO
- Device is maintained in an open position and has to be closed voluntarily by pulling with the cable on the harness system to grasp an object.
- To release, the patient releases the pull on the harness, and a spring in the terminal device opens it.
- The maximum prehensile force is determined by the strength of the individual. *Disadvantages:* Prolonged prehension requires constant pull on the harness; the terminal device remains awkwardly open when not in use.

3. **Externally powered (electric-powered) terminal devices**:
   - Controlled by switches or myoelectric signals and are powered with energy provided from external batteries
   - The electric-powered terminal device can be hand-like (e.g., Ottobock System Electric Hands or Steeper Electric Hands), nonhand-like (Ottobock Greifer Hand, Hosmer NU-VA synergistic prehensor, and Steeper Powered Gripper), or hook-shaped in appearance.
   - The second generation of electric-powered hands (smart hands) provides multiple prehension modes including grasp, pinch, point, peck, and others (I-Limb Hand, Bebionic Hand, Michelangelo Hand; manufacturer websites: touchbionics.com; bebionic.com; ottobockus.com).

   a. *Myoelectric-controlled terminal devices:*
   - Use surface electrodes placed on the muscles of the residual limb
   - Devices can have a digital or proportional control system:
     - Digital control system: On/off system
     - Proportional control system: The stronger the muscle contraction producing the signal, the faster the action.

   b. *Microswitch-controlled terminal devices:* Use either a push-button switch or a pull-switch to activate the TD

🕮 PROSTHETIC WRIST UNITS

- Wrist units are used for attaching terminal devices to prostheses as well as providing pronation and supination to place the terminal device in its proper position.
- The rotation function is passive: The amputee rotates the terminal device in the wrist unit with their sound hand or by pushing against a part of the body or other surface to produce either pronation or supination. The wrist unit also permits interchange of the terminal devices.
- 🕮 The wrist flexion unit allows the terminal device to be in the flexed position, facilitating ability to perform activities close to the body. This is important for bilateral upper extremity amputees (need flexion for activities of daily living [ADLs]: Feeding, wiping; if unilateral, upper extremity amputee can use other hand).
- 🕮 Electric wrist rotator units are also available and are generally considered for bilateral upper extremity amputees.
- Do not add components unnecessarily: They increase weight and cost, and can break and need repair.
- Two types of wrist units: *Friction control* or *locking*

1. *Friction wrists* permit pronation and supination of the terminal device and hold it in a selected position by means of friction derived from a compressed rubber washer or from forces applied to the stud of the terminal device.

2. *Locking wrists* permit manual rotation and then lock the terminal device in its fixed position.
   - *Advantage:* The locking mechanism prevents inadvertent rotation of the terminal device in the wrist unit when a heavy object is grasped.

- Two types of wrist flexion units: *Add-on* and *combination*
  - *Add-on* is worn between the wrist and the terminal device and allows manual positioning of the terminal device in either the straight or the flexed position.
  - *Combination type* combines a friction wrist and a wrist flexion component in one and provides for setting and locking in one position.

Transradial (Below-Elbow) Amputation Prostheses

- In addition to terminal devices and wrist units (discussed previously), a transradial amputee with a body-powered prosthesis will also need the following:
  1. Socket
  2. Elbow hinge
  3. Upper arm cuff/pad
  4. Harness and control system

SOCKETS FOR TRANSRADIAL AMPUTATIONS

- The socket must provide a comfortable but stable, total-contact interface with the residual limb (to avoid inadvertent motion and prevent uncomfortable concentrations of pressure), efficient energy transfer from the residual limb to the prosthetic device, secure suspension, and adequate appearance.
- To accomplish these goals, most sockets are double-walled with the inner wall giving total-contact fit and the outer wall matching the contour and the length of the contralateral forearm.
- Proximally, the socket extends posteriorly to the olecranon and anteriorly to the elbow crease. The shorter the residual limb, the closer is the trim line to the crease.
- Prosthetic socks or a soft interface material may be used to make the socket fit comfortably and to allow for some accommodation of volume changes.
- Special socket designs
  1. **Split socket:**
     - Split socket consists of a total-contact segment encasing the residual limb that is connected by hinges to a separate forearm shell to which the wrist unit and the TD are attached.
     - It is sometimes used in *patients who have very short residual limbs* so that the special elbow hinges can be used to increase available joint ROM or to incorporate an elbow-lock mechanism in the prosthesis.
  2. 🕮 **Muenster socket (self-suspended socket; Figure 6–5):**
     - An alternative to the split socket (for short transradial amputees) is the self-suspended Muenster-type transradial socket design.
     - In this, the socket and forearm are set in a position of initial flexion and the socket encloses the olecranon and the epicondyle of the humerus. The intimate residual limb encapsulation, flexion attitude, and high trim lines provide suspension. Although there is some limitation in flexion-extension ROM, this *is compensated by preflexing the socket*.
     - When this type of suspension is used, a figure-9 harness can be used for control purposes only.
     - With these prostheses, the patient can operate the TD in common positions and still apply full torque about the elbow. Although these techniques yield less elbow flexion than the split socket, the reduction in force requirements and the ease of use more than compensate for this limitation.

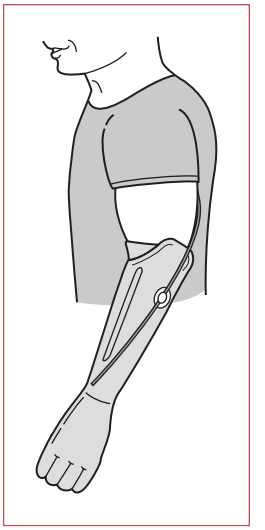


FIGURE 6–5 The Muenster socket.

ELBOW HINGES

- Transradial amputation prosthesis elbow hinges connect the socket to a cuff on the upper arm and are important for suspension and stability.
- Types:
  1. **Flexible elbow hinge:**
     - Used primarily to suspend the forearm socket
     - Permits active pronation and supination of the forearm
     - Used where sufficient voluntary pronation and supination are available to make it desirable to maintain these functions. Wrist disarticulations and long transradial amputations are common.
  2. **Rigid elbow hinge:**
     - Used in short transradial amputations when normal elbow flexion is present but there is no voluntary pronation-supination and more stability is needed
     - Types:
       a. Single axis
       b. Polycentric
       c. Step-up design—used with split-socket prosthesis, in very short transradial amputation where flexion is limited. By virtue of a gear or double-pivot arrangement, these hinges permit the residual limb to drive the prosthetic forearm through an increased ROM. The disadvantage is that it takes twice the force (energy cost doubles) to provide the same amount of flexion as a single-axis hinge joint.
  3. **Locking elbow joints** are used only if there is significant elbow flexor weakness.

CUFFS AND PADS

- Except in the Muenster socket, a half-cuff or a triceps pad with an appropriate elbow hinge is used on the upper arm to connect the socket to the harness and help furnish socket suspension and stability.
- It also serves as an anchor for the cable-control action point. The half-cuff is used in the majority of short transradial fittings. The triceps pad is used with long transradial, wrist disarticulation, and transmetacarpal prostheses.

TRANSRADIAL HARNESS SUSPENSION SYSTEMS

- The functions of the below-elbow harness are to suspend the prosthesis from the shoulder so the socket is held firmly on the residual limb, utilize body motions as sources of power or force, and transmit this force via a cable system to operate the TD.
- Three types:
  1. 🕮 **Figure-8 (O-ring) harness (Figure 6–6):**
     - Most commonly used harness. The axilla loop, worn on the sound side, acts as a reaction point for the transmission of body force to the TD.

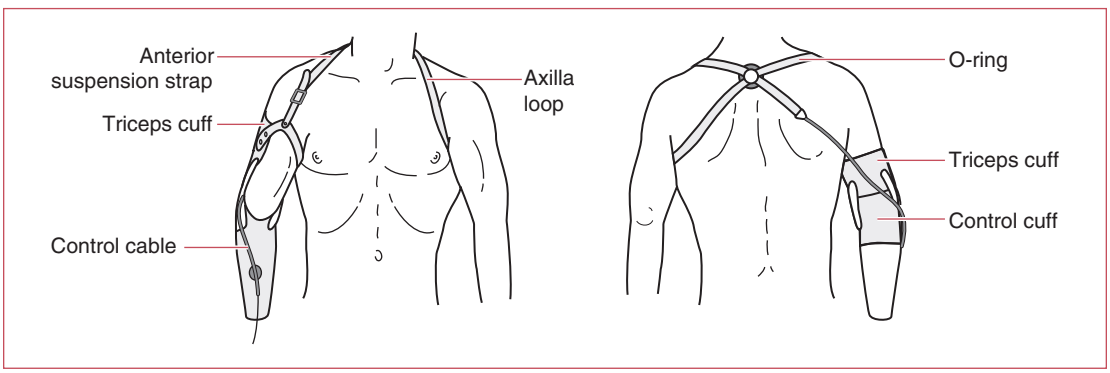


FIGURE 6–6 O-ring (figure-8) harness.

- The anterior suspension strap on the involved side gives additional support for pulling or lifting, and acts as the attachment point for the elbow loading strap on a body-powered above-elbow prosthesis.

2. **Figure-9 harness:**
   - A figure-9 harness is often employed with a self-suspended transradial socket (e.g., Muenster socket) that requires a harness only for controlling the TD or for long residual limbs with suction sockets.
   - Consists of an axilla loop and a control attachment strap
   - *Pros*: It is lighter and provides greater freedom and comfort by elimination of the usual front support strap and triceps pad or cuff.
3. **Chest strap with shoulder saddle (Figure 6–7):**
   - Chest strap with shoulder saddle is used if the patient cannot tolerate the axilla loop. Also used with those who will be doing heavy lifting.

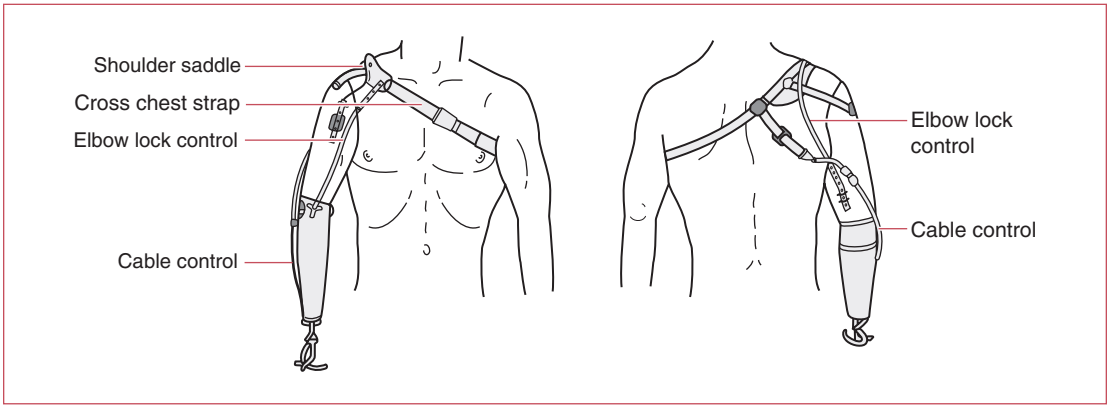


FIGURE 6–7 Shoulder saddle harness (modified) for above-elbow prosthesis.

CONTROL CABLE SYSTEMS

- The typical control cable system for transmission of power to the prosthesis consists of a flexible, stranded stainless steel cable which slides within a flexible housing. The cable is attached proximally at the harness and distally at the elbow or TD.
- There are two types of cable systems:
  1. The **single-control cable system** (Bowden cable system) is used in the transradial single-control cable system and consists of one cable to transmit body power for the single purpose of operating the TD.
     - Consists of a continuous length of flexible housing through which the cable slides. The housing is fastened by a base plate and retainer to the forearm shell and by a housing crossbar assembly to the cuff of the triceps pad; these housing retainers also serve as reaction points when force is applied to the cable.
     - The muscle movements used to operate the terminal device are forward humeral flexion and biscapular abduction.
  2. The **dual-control cable system** (split cable or fair-lead cable system) is typically used in the transhumeral control cable system as well as in the very short transradial split-socket prosthesis with locking hinge.
     - Consists of one cable with two functions:
       a. *Flex* the elbow unit when the elbow is unlocked
       b. *Operate* the terminal device when the elbow is locked
     - The cable is held in place and guided by separate lengths of housing. The pieces of housing are fastened with retainers at points where the cable must be supported or operated through an angle.

- Since the system must provide force for elbow flexion and operation of the terminal device, two fair-lead housings are necessary:
  - The proximal lead, through which the cable slides when the elbow is flexed
  - The distal lead, through which the cable slides when the terminal device is operated

Elbow Disarticulation Prostheses

- Variation of a transhumeral prosthesis
- Socket is flat and broad distally to conform to the epicondyles of the distal humerus, which provides self-suspension and allows for internal and external rotation of the humerus.
- Length of residual limb requires use of external elbow joint with cable-operated locking mechanism.
- Harness and control system are the same as for transhumeral prostheses.

Transhumeral (Above-Elbow) Prostheses

- In addition to the terminal device and wrist unit, a transhumeral body-powered prosthesis consists of:
  - Forearm
  - Elbow unit
  - An upper arm
  - Socket
  - Harness and control cable system
- Terminal devices and wrist units are the same as those used for transradial prostheses, but the socket, elbow unit, harness, and control systems differ in several respects from those used in transradial prostheses.

SOCKETS FOR TRANSHUMERAL AMPUTATIONS

- As in transradial prostheses, the transhumeral socket is usually of double-wall construction, with the inner wall providing a snug, total-contact fit and the outer shell providing appropriate length and shape. The lateral socket wall extends to the acromion and the medial socket wall is flattened below the axilla to help prevent inadvertent socket rotation. A soft interface material is often used to make the socket fit more comfortably.

ELBOW UNITS

- When an amputation occurs at or above the elbow joint, elbow function is supplied by the use of an elbow unit, which provides for elbow flexion and for locking in various degrees of flexion.
- Elbow-locking systems are divided into two types:
  1. *External (outside) locking elbow*: Used with elbow disarticulations because there is not enough space for an internal locking mechanism
  2. *Internal locking elbow*: Used with transhumeral and shoulder prostheses
- *Internal elbow joint:*
  - Preferred because of greater mechanical durability and is cosmetically more appealing
  - Used in level of amputation 4 cm or more proximal to the level of the epicondyles
- *External elbow joint:*
  - Used when the residual limb extends more distally than 4 cm to the level of the epicondyles to maintain the elbow joint center equal to that in the nonamputated side
- Both types of elbows are flexed by the dual-control cable system and locked at the desired flexion angle by a separate elbow-lock control cable, which is attached at one end to the elbow mechanism and at the other end to the anterior suspension strap.
- The locking mechanism operates on the alternator principle: Locking and unlocking actions alternate with each control cable cycle of tension and relaxation.
- For amputees who have difficulty flexing their prosthetic forearm, an accessory in the form of a spring assist for elbow flexion may be provided for use with the internal elbow.

- In transhumeral and shoulder prostheses, passive humeral rotation is accomplished by means of a turntable between the elbow unit and the upper arm shell or socket. As in the case of a wrist unit, friction between the elbow unit and the turntable permits control of the rotation to maintain the desired plane of elbow operation.

TRANSHUMERAL HARNESS SUSPENSION AND CONTROL SYSTEMS

- **Transhumeral harness**:
  - In addition to suspending the prosthesis from the shoulders, the transhumeral harness must transmit power to flex the prosthetic forearm, to lock and unlock the elbow unit, and to operate the terminal device.
  - 🕮 The harness designs *most frequently used* for transhumeral prostheses are modifications of the basic figure-8 and chest strap patterns used with transradial prostheses.
- **Body-powered control cable system** in transhumeral prostheses:
  - As previously mentioned, the *dual-control (fair-lead control) cable system* is used in the transhumeral prosthesis to transmit force for two functions:
    1. Elbow flexion
    2. Terminal device operation
  - *Elbow locking and unlocking* are controlled by a second cable, the elbow-lock cable.
  - When the elbow is extended and unlocked, *flexing the shoulder* (humeral flexion; assisted by biscapular abduction) transmits force to the forearm lever loop *flexing the elbow to the desired level*.
  - If the amputee wishes to use the terminal device at this point, first, they lock the elbow (by doing shoulder depression, extension, and abduction ("down, back, out").
  - Then, the patient can operate the terminal device by continuation of the control motion ⇒ shoulder flexion and biscapular abduction.
  - The same combination of shoulder movements is done to lock or unlock the elbow (shoulder depression, extension, and abduction). The elbow extends by gravity when unlocked.

🕮 DUAL-CONTROL CABLE MECHANISM OPERATION IN THE TRANSHUMERAL AMPUTEE (TABLE 6–7)

Myoelectric Control Systems

- Externally powered systems for below-elbow and above-elbow amputees
- Myoelectric control relies on activation of muscles in the residual limb or proximal segment.
- The electrical signal produced by voluntary activation of the residual muscles is detected by surface electrodes incorporated into the prosthetic socket.

TABLE 6–7 Dual-Control Cable Mechanism Operation in the Transhumeral Amputee

	1. ELBOW FLEXION	2. LOCK ELBOW (IN DESIRED DEGREE OF FLEXION)	3. TERMINAL DEVICE OPERATION (OPENING AND CLOSING)	4. UNLOCK ELBOW
Movements	Humeral flexion and biscapular abduction	Shoulder depression, extension, and abduction (down, back, out)	Further humeral flexion and biscapular abduction	Shoulder depression, extension, and abduction (down, back, out)
Cable mechanism	Dual-control cable	Single-control cable (elbow-lock cable)	Dual-control cable	Single-control cable (elbow-lock cable) After desired terminal device function is accomplished, the elbow is unlocked and the elbow extends by gravity

- In below-elbow amputees, wrist extensors (extensor carpi radialis longus/brevis and extensor carpi ulnaris) are used to open the terminal device, and wrist flexors (flexor carpi radialis and flexor carpi ulnaris) are used to close the terminal device.
- In an above-elbow amputee, the biceps muscle is used to flex the elbow and close the terminal device, while the triceps muscle is used to extend the elbow and open the terminal device. The patient can change back and forth between the terminal device and the elbow control by co-contracting the biceps and triceps. With short transhumeral or shoulder amputation, shoulder girdle muscles are used to control elbow function and terminal device function.
- Hybrid control systems may combine body-powered and myoelectric control in the above-elbow or shoulder prosthesis. This can reduce the cost, weight, and harnessing of the prosthesis.
- Electric switches can also be incorporated into the harness or socket if myoelectric control is not available.

🕮 BODY-POWERED VERSUS MYOELECTRIC CONTROL SYSTEMS (TABLE 6–8)

- **Body-powered devices:**
  - *Advantages:* Less expensive, lighter, more durable, easier to repair, higher sensory feedback
  - *Disadvantages:* Mechanical appearance, difficult to use for some people, dependent on motor strength
- **Myoelectric devices:**
  - *Advantages:* Better cosmesis, less harnessing, stronger grasp force
  - *Disadvantages:* More expensive, heavier, decreased durability due to electronic components and the need for daily recharging of batteries

TABLE 6–8 Advantages and Disadvantages of Body-Powered and Myoelectric Devices

	BODY-POWERED DEVICES	MYOELECTRIC DEVICES
Advantages	• Lower initial cost • Lighter • Easier to repair • Offer better tension feedback to the body	• Do not require a harness or cable • Looks like natural-appearing arm • Battery-powered, so motor strength and coordinated mobility not as important • Newer batteries have reduced weight • Provides strong grip force
Disadvantages	• Dependent on motor strength • Mechanical appearance • Some people have difficulty using them	• Higher initial cost • Heavier • Dependence on battery capacity, voltage, and life • Higher repair cost

Source: From Braddom RL, Chan L, Harrast MA. *Physical Medicine and Rehabilitation*. Saunders/Elsevier; 2015.

Shoulder Prostheses

- All shoulder prostheses consist of a terminal device, wrist unit, forearm section, elbow unit, humeral and shoulder sections, harness, and control cable system.
- Terminal device, wrist unit, forearm section, and elbow units are identical to those used in transhumeral prostheses.
- The shoulder section includes a socket, which provides:
  - Comfortable, stable bearing on the residual shoulder elements and thorax
  - Means of utilizing the remaining shoulder girdle mobility for control of the prosthesis

ISSUES IN UPPER EXTREMITY AMPUTEE CARE AND REHABILITATION

- There are three important considerations in the clinical decision-making process from a medical standpoint for a prosthesis:
  1. Determination of amputation level

2. Careful assessment of bilateral proximal muscle strength and ROM when planning for prosthetic control mechanisms
3. Evaluation of general health and cognition

- In upper extremity amputations, general health is typically no different than in the regular population. *Exception:* Those with malignancies.
- Assess for conditions that should be treated before the patient is fitted with a prosthesis. Cognitive impairment and other neurologic problems can hinder prosthetic training.
- **Diagnosis of specific residual limb conditions:**
  - Abnormalities such as bone spurs, tender scars, skin rashes, neuromas, and other conditions are of low statistical importance.
  - When present, many of these can be dealt with by suitable fitting modifications.

Other Rehabilitation Issues

- **Preprosthetic training:**
  - Another important aspect of the initial management of the upper limb amputee is preprosthetic therapy with physical therapy and occupational therapy.
  - *Preprosthetic training includes* residual limb shrinkage, muscle strength and ROM, postural problems, desensitization, scar mobilization, ADL assessment, and home exercise program.
  - Exercises should be prescribed if there are any deficiencies. Forearm and humeral rotations are the motions most seriously affected.
    - Transhumeral amputations: 80% have limitations of humeral rotation.
    - Transradial amputations: 80% have limitations of forearm rotation.
  - If bony malformations limit ROM, physical therapy will be of no help.
  - Testing and strengthening muscles in the residual limb may be helpful for future myoelectric control.
- **Vocational issues:**
  - The terminal device is extremely important to the amputee's vocational success.
  - The greatest occupational effect of amputation is on skilled and semiskilled laborers who are unable to continue their original occupations. This is a source of emotional maladjustment and of reduction in earning power.
  - The prosthetic team should serve in an advisory capacity on any contemplated change in occupation because the patient may be able to return to their job with the prosthesis.
- **Emotional/psychological issues:**
  - Although there is no evidence to indicate that the amputee population varies from the general population with regard to emotional stability, psychological problems can act as a deterrent to rehabilitation. It is a waste of time and money to fit an amputee with a prosthesis and then to accept failure because the amputee does not cooperate fully. There is always a period of adjustment to limb loss. Psychological counseling should be made available to all amputees to deal with grieving, anger, and depression.
  - Personal factors also must be taken into consideration. One should consider age, sex, educational history, prosthetics history, and personal preference. In many cases, the educational background completes the picture of the amputee and gives us a better idea of the prescription necessary. Most do not have personal preferences for the prosthesis, but if they exist, they must be explored. If these preferences cannot be accommodated, the reason must be explained to the amputee.

Prosthetic Operation and Training

- In the unilateral amputee, the prosthesis generally will be used to assist the remaining limb, allowing a portion of the lost function to be regained. The prosthetic device is still far from duplicating the lost part and the patient must be aware of its functional limitations.
- The training period consists of an orientation to the prosthesis, its controls, and use of the prosthesis, which initially emphasizes ADLs.

- Introduction to the prosthesis from the preprosthetic phase should be repeated and the patient should be specifically instructed in the correct terminology and function of each component. This understanding will help the patient communicate with the prosthetist if something is wrong. General instruction about the care of the prosthesis should be covered at this time.
- The next step for training is instruction for donning and doffing the prosthesis. After this initial instruction, the patient is ready to begin learning the basic motions of operation. The therapist should take the amputee through each required motion so that the amputee can see and feel the motion being performed. The patient then repeats the same motion independently.

TRANSRADIAL AMPUTEE TRAINING

- Training is concentrated on operation of the terminal device.
- Forearm and elbow control requires no special training.
- 🕮 Myoelectric transradial prostheses use muscle contractions to activate the prosthesis (e.g., activation of wrist flexors close terminal device and activation of wrist extensors open terminal device).
- Body-powered prostheses use forward flexion of the humerus with some assistance from biscapular abduction to open the terminal device. The shoulder on the amputated side should not flex excessively to give a smooth motion. The shoulder on the opposite side acts as a stabilizer.
- Once the patient has learned the mechanics of the prosthesis and how to use it efficiently, they are ready for training in purposeful activity. The therapist should present different activities to help solve new problems that inevitably arise in the patient's life. Before attempting any activity, prepositioning of the terminal device is essential. Instruction and practice in this prepositioning are necessary. It will allow an amputee to approach an object correctly.
- Prehension is the final phase of control training before practice in daily activities is started. Drills in the approach, grasp, and release of various sizes of objects and different types of materials are used. The amputee is taught to grasp objects with adequate pressure control of the terminal device.
- Control training may be considered complete when the patient has maximal control of the terminal device in space.
- Once basic operations are learned, these techniques are applied to practice ADLs. The amputee should gain confidence in using the prosthesis in a wide range of activities. Initially, the activities of most importance for the amputee are feeding and dressing.
- For the unilateral amputee, this independence is not difficult to achieve. A prosthesis is not needed to achieve basic independence. Activities chosen should require the use of two hands (e.g., cutting food with a knife and fork or tying shoelaces). As the patient attempts, performs, and succeeds in these activities, they become more willing to accept the use of the prosthesis and can rely on it.
- After training in feeding, dressing, and grooming is completed, specialized activities such as communication skills involving the use of the telephone or keyboard can be addressed. Homemaking, vocational, and recreational interests should be encouraged, and the activities associated with these interests should be emphasized in the training process.

ELBOW DISARTICULATION AND TRANSHUMERAL AMPUTEE TRAINING

- The prosthetic training for the elbow disarticulation or transhumeral amputee follows the same general principles as for the transradial amputee.
- Control training of this level of amputee is more difficult since the amputee must now concentrate on locking the prosthetic elbow before being able to use the terminal device.
- Training with dual-control/elbow-locking transhumeral prosthesis should not be attempted prior to age 3.
- When the elbow is unlocked, humeral flexion produces flexion of the forearm section of the prosthesis (elbow flexion).

- The shoulder on the amputated side should not flex more than is necessary for smooth movement. The opposite shoulder acts as a stabilizer. This should be repeated by the amputee until the speed of the movement and the angle of flexion are smooth and controlled.
- Elbow extension (with elbow still unlocked) is achieved by slowly bringing the shoulder back to its starting position.
- Next control motion is for locking and unlocking the elbow:
  - This motion is a combination of shoulder depression, extension, and abduction.
  - This motion should be practiced with the elbow extended until the lock clicks.
  - Once the locking and unlocking of the elbow can be accomplished smoothly with the elbow extended, the amputee can be taught to flex the elbow and maintain tension on the cable while the elbow lock is used.
- To unlock the elbow, the amputee repeats this procedure, allowing the forearm to return smoothly to the starting position.
- When the elbow is locked, additional humeral flexion will operate the terminal device (opening in VO terminal device or closing in VC terminal device).
- For the terminal device to be operated closer to the body, rotation of the prosthetic forearm is necessary. To do this, the amputee is instructed to first flex the elbow to 90 degrees and then manually rotate the turntable medially or laterally.

LOWER LIMB AMPUTATION AND PROSTHETICS

Lower Limb Amputations

- From 1989 to 1992, there were approximately 105,000 amputations of the LE; 32% of those were toe amputations (Seymour, 2002).

AGE GROUP (YEARS)	MOST COMMON CAUSES OF AMPUTATION
0–5	Congenital or deformity requiring amputation
5–15	Cancer, trauma
15–50	Trauma
50+	Vascular disease, infection

Distribution of Lower Extremity Amputation and Cause in Survey by Kay and Newman of 5,830 New Amputees

DISTRIBUTION	%
Syme's	3
Transtibial	59
Knee disarticulation	1
Transfemoral	35
Hip disarticulation	2

CAUSES	%
Peripheral vascular disease and infection	70
Trauma	22
Tumor	5
Congenital deformity	3

Source: From Kay HW, Newman JD. Relative incidences of new amputations: statistical comparisons of 6,000 new amputees. *Orthot Prosthet*. 1975;29:3–16. http://www.oandplibrary.org/op/1975_02_003.asp, with permission.

🕮 DIAGNOSTIC TESTING FOR PERIPHERAL ARTERIAL DISEASE

- PAD, also referred to as PVD, is one of the most common causes of LE amputations, especially in people ages 50 and older (Savji et al., 2013). Appropriate vascular screening should be performed in patients who are at risk.

- **Ankle–brachial index:**
  - PAD is usually diagnosed with ABI, which is the ratio of brachial systolic pressure to ankle systolic pressure.
  - Economical and easy to perform, but accuracy is limited especially for large vessels and for calcified vessels, which can occur in patients with diabetes, elderly, or renal failure patients (DeLisa et al., 2005)
  - Not a reliable tool for determining the level of an amputation
  - Ranges (Andrews et al., 2019):

ABI RANGE	DESCRIPTION
1.0–1.39	Normal
0.9–0.99	Borderline
0.8–0.89	Mild PAD
0.5–0.79	Moderate PAD
0.0–0.49	Severe PAD
≥1.40	May suggest calcified, noncompressible vessels, which can produce false negative results. This is common in patients with diabetes.

ABI, ankle–brachial index; PAD, peripheral arterial disease.

- Mild to moderate PAD patients likely experience pain with exercise (claudication; Khan et al., 2008).
- Severe PAD patients experience pain while resting, with increased risk of gangrene, ulceration, and impaired wound healing (Khan et al., 2008).
- 🕮 Poorly compressible arteries (ABI ≥1.4) are associated with limb amputation, while severe PAD (ABI ≤0.5) was associated with limb revascularization procedure (Moussa Pacha et al., 2018).
- **Doppler velocity waveform analysis:**
  - If screening ABI is abnormal, Doppler waveform analysis is performed to localize the lesion.
  - Doppler waveforms are obtained at multiple sites, and a change in waveform from one level to the next is indicative of PAD.
- **Segmental pressure analysis:**
  - Measurements of segmental pressures via cuffs or Doppler can help localize the location of arterial stenosis in PAD (Andrews et al., 2019).
  - Presence of a significant systolic pressure gradient (greater than 10–15) between segmental pressures at two locations suggests obstruction somewhere in between (e.g., significant systolic pressure gradient between upper and lower thigh pressures suggests superficial femoral artery stenosis; Andrews et al., 2019).
  - 🕮 Preoperative lower limb segmental blood pressures may predict healing postamputation and guide level of amputation. Lepäntalo et al. found postoperative healing occurred in all studied cases when preoperative calf systolic pressures were 68 mmHg or more and distal thigh systolic pressures were 100 mmHg or more. When calf pressures were below 35 mmHg and distal thigh pressures were 60 mmHg or less, all amputations failed (Lepäntalo et al., 1982).
- **Intra-arterial contrast angiography:**
  - Gold standard imaging test for PAD
  - Invasive test and typically not used for screening purposes

IMPORTANT DEFINITIONS

- 🕮 **Myodesis**: Muscles and fasciae are sutured directly to bone through drill holes.
  - Residual limb is more structurally stable.

  - Contraindicated in severe dysvascularity in which the blood supply to the bone may be compromised
- 🕮 **Myoplasty:** Opposing muscles are sutured to each other and to the periosteum at the end of the cut bone with minimal tension.
  - Generally takes less operating time
  - May be the procedure of choice in severely dysvascular residual limbs

Levels of Amputation in the Lower Limb (Figure 6–8; Table 6–9)

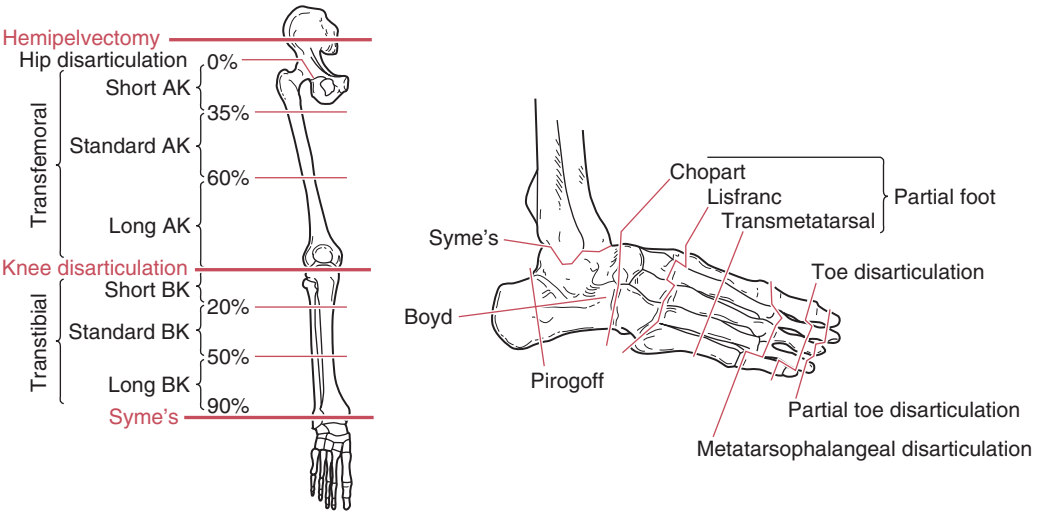


FIGURE 6–8 Levels of lower extremity amputations.
AK, above knee; BK, below knee.

TABLE 6–9 Descriptions of Level of Amputation in the Lower Limb

Partial toe	Excision of any part of one or more toes
Toe disarticulation	Disarticulation at the MTP joint
Partial foot/ray resection	Resection of a portion of up to three metatarsals and digits
TMA	Amputation through the midsection of all metatarsals
Lisfranc	Amputation at the tarsometatarsal junction
Chopart	Midtarsal amputation—only talus and calcaneus remain
Syme's	Ankle disarticulation with attachment of heel pad to distal end of tibia; may include removal of malleoli and distal tibial/fibular flares
Transtibial: Long BKA → Standard BKA → Short BKA →	 >50% of tibial length 20%–50% of tibial length <20% of tibial length
Knee disarticulation	Amputation through the knee joint, femur intact

(*continued*)

TABLE 6–9 Descriptions of Level of Amputation in the Lower Limb (*continued*)

Transfemoral: Long AKA ⟶ Standard AKA ⟶ Short AKA ⟶	 >60% of femoral length 35%–60% of femoral length <35% of femoral length
Hip disarticulation	Amputation through hip joint, pelvis intact
Hemipelvectomy	Resection of lower half of the pelvis
Hemicorporectomy	Amputation of both lower limbs and pelvis below L4–L5 level

AKA, above-the-knee amputation; BKA, below-the-knee amputation; MTP, metatarsophalangeal; TMA, transmetatarsal amputation.

Indications, Advantages, and Disadvantages of Common Levels of Amputation in Lower Extremities

UNSATISFACTORY LEVELS FOR ELECTIVE SITES OF LOWER LIMB AMPUTATION

- Distal two-fifths of tibia (below gastroc-soleus musculature):
  - The added length of the lever arm at this level is outweighed in importance by difficulties of good skin and soft tissue management.
  - Because the tibia and fibula are both subcutaneous at this level, there is inadequate muscle coverage for the distal tibia.
- Very short below-the-knee amputation (BKA) proximal to the tibial tubercle:
  - Knee extension strength is lost, and the knee becomes valueless for stability and ambulation.
  - The added length creates difficulties in prosthetic fit as compared with a knee disarticulation. Knee flexion contractures occur often.
  - When active knee extension is present and the knee can be flexed through a range of 45 degrees or more, the residual limb can be comfortably fit with an effective prosthesis if surgery is performed at least 1 cm below the tibial tubercle. Above this level, most patients will be better served by a knee disarticulation.
- Very high above-the-knee amputation (AKA)
  - When transection is just a short distance below the lesser trochanter, the limb tends to develop excessive flexion and abduction at the hip joint.
  - Socket fit may become a difficult problem. Releasing the contractive deformity through flexor and abductor muscle release at the hip can circumvent this. Many surgeons prefer to leave a short segment of femur rather than amputate at the hip disarticulation level.

Foot and Ankle Amputations (Figure 6–9)

PARTIAL TOE AMPUTATION, TOE DISARTICULATIONS, AND METATARSAL RAY RESECTION

- Performed for deformity resulting from trauma to the toes, loss of tissue, infection, gangrene due to frostbite, diabetes, arterial sclerosis, scleroderma, Buerger's disease, and similar conditions
- By preserving the length of the foot, there is a better mechanical advantage.
- There is better gait pattern compared with a transmetatarsal amputation (TMA).
- *Disadvantages:* Mostly seen in the vascular patient, including arterial sclerosis and diabetes. One must be assured that the vascular supply to the rest of the foot will be intact so as not to have a continuous progression of amputations up to the transtibial level.

TRANSMETATARSAL AMPUTATION

- Also performed for deformity resulting from trauma to the toes, loss of tissue due to an infection, or gangrene due to frostbite, diabetes, arterial sclerosis, scleroderma, Buerger's disease, and similar conditions.

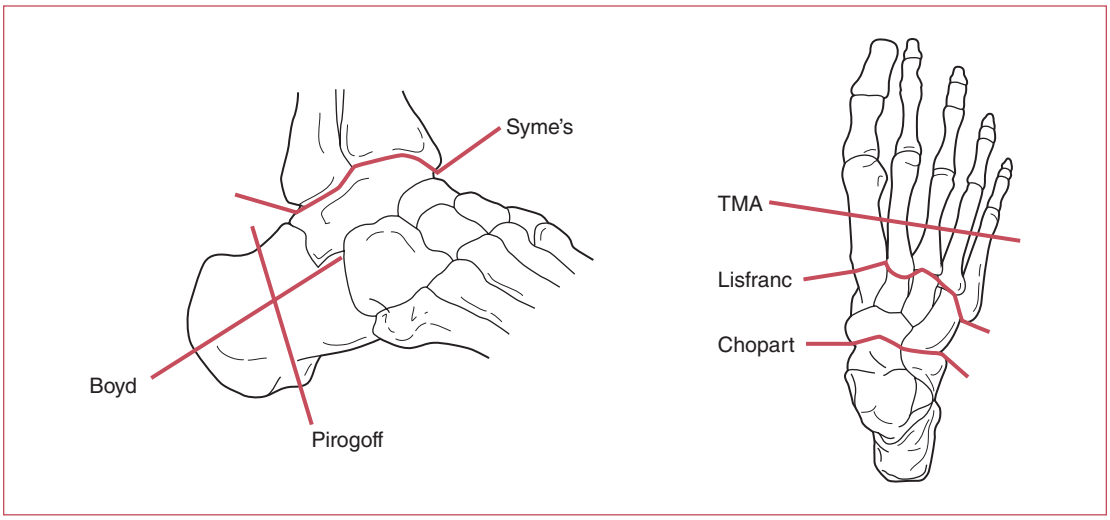


FIGURE 6–9 Foot and ankle amputations.
TMA, transmetatarsal amputation.

1. Gangrene must be limited to the toes and should not involve the web space. Infection should be controlled. The incision should not extend through hypesthetic areas or through infected areas. The patient should be free of pain. Palpable foot pulses are not necessary, but there should be no dependent rubor. Venous filling should be <25 seconds.
2. Not indicated in cancers of the metatarsal bones because it would not be conclusive that all bony cancer would have been resected.
3. This is an important amputation through the foot because it preserves the attachment of the dorsiflexors and plantar flexors and their function. This continues to give the patient a good mechanical advantage with the foot.
4. For functional use, it is also a good level of amputation because these amputations can be fitted with sole stiffeners and toe fillers with minor apparent loss of function during stance and walking on level surfaces.

🕮 SYME'S AMPUTATION

- It is essentially an ankle disarticulation procedure with attachment of the distal heel pad to the end of the tibia; may include removal of the malleoli and/or distal tibial/fibular flares.
  - *Indications:* Trauma of the foot, congenital anomalies, tumors, and deformities that necessitate amputation
  - *Disadvantages:* Healthy plantar heel skin is necessary for weight bearing in this area. The patient also must have good perfusion in this area, thus making it a difficult procedure for the dysvascular patient.
  - *Pros:* Functionally, this procedure represents an excellent level of amputation because:

1. It maintains the length of the limb.
2. There is preservation of the heel pad, providing an excellent weight-bearing residual limb.
3. Early fitting of prosthesis is possible with excellent results.
4. Partial weight bearing of the residual limb is possible almost immediately after the procedure with a proper rigid casting (approximately within 24 hours).

- *Cons*: ↓ cosmesis (bulbous, bulky residual limb); fitting for a prosthesis may be more difficult than for other amputation levels.

OTHER FOOT AMPUTATIONS

- 🕮 **Lisfranc** amputation at the tarsometatarsal junction
- 🕮 **Chopart** amputation removes all the tarsals and the metatarsals. Only talus and calcaneus bones remain.

- A **Pirogoff** amputation is a vertical calcaneal amputation.
- A **Boyd** amputation is a horizontal calcaneal amputation:
  - These amputations are not indicated in patients with ischemia or diabetes. They may be indicated, at times, in patients with trauma, although their outcomes are still poor.
  - 🕮 In both the Lisfranc and Chopart amputations, the remaining foot often develops a significant equinovarus deformity resulting in excessive anterior weight bearing with breakdown. Adequate dorsiflexor tendon reattachment with Achilles tendon lengthening has been advocated to prevent this deformity.

Transtibial Amputation/Below-the-Knee Amputation

- When an amputation in the lower limb becomes necessary due to trauma, tumor, or disease, the importance of saving the knee joint cannot be overemphasized. Preservation of the knee joint means preservation of a near-normal lifestyle with minimal physical limitations.
- In the elderly amputee, saving the knee may well mean the difference between being able to walk and being confined to a wheelchair.
- 🕮 In the elderly, approximately 50% of patients have worse functional ability after a BKA. Approximately 5% of patients actually improve their functional ability.
- Studies performed to compare the energy requirements of below-the-knee amputees with above-the-knee amputees in walking have consistently shown significantly greater demands at the above-the-knee level.
- The ability of performing a BKA as opposed to a higher level amputation will be determined by tissue viability in cases of trauma and by principles of tumor surgery when dealing with neoplasm.
- In the case of the dysvascular patient, BKA is not only feasible, it is the level of choice in most instances.
- Reported healing rate for ischemic BKAs is 80% to 90%.
- There is also general agreement in the literature that mortality is lower following BKAs than after AKAs because BKAs have higher healing rates and better tissue viability and weight-bearing surfaces. BKAs also show decreased mortality and decreased energy expenditure.
- BKA should be in the proximal 50% of the tibia. Within this range, preserving length is critical as long as viable skin and muscle is available for distal coverage. The longer the below-the-knee residual limb, the greater will be its leverage, strength, and proprioceptive qualities, and the better the amputee will walk. The energy costs of walking will be less if a longer amputation residual limb is provided to the amputee. On the other hand, a very short below-the-knee residual limb is far superior to a knee disarticulation, and satisfactory fitting with a below-the-knee prosthesis can be accomplished at levels as high as the tibial tuberosity.
- The fibula should be cut 2 to 3 cm shorter than the tibia, and the tibia should be beveled anteriorly.
  - An alternative surgical procedure is the Ertl procedure creating a bone bridge/fusion between the tibia and fibula. The advantage is potential for end-bearing in the prosthesis.
- If there is a severe flexion contracture (>50 degrees) and the limb is ischemic, knee disarticulation should be the procedure of choice, although a very short BKA may be fitted as a knee disarticulation with a bent-knee prosthesis.

FUNCTIONAL OUTCOMES AFTER BELOW-THE-KNEE AMPUTATION

- Preoperatively, the patient should be familiarized with the procedures and activities to be performed after the amputation. Instruction should be given in the following:
  1. Proper bed and wheelchair positioning
  2. Exercises to be executed, as well as the rationale for such treatment
  3. If possible, the patient should be taught ambulation techniques with appropriate assistive devices.

4. The type of prosthetic device to be used postoperatively; in using a below-the-knee prosthesis, there is much less energy expenditure, and the patient's functional abilities should be greater than those of a patient with an AKA.
5. Prolonged sitting after the amputation should be avoided to prevent contractures. The increase of contractures will result in a decrease in the patient's functional ability.

Knee Disarticulation

- *Advantages:*
  - Less traumatic to tissue
  - Blood loss is minimal.
  - A long, strong residual limb with excellent end-bearing quality is produced.
  - Prosthetic suspension is facilitated by the bulbous contour of the residual limb end.
- *Disadvantages:*
  - Relatively long flaps are necessary and healing may be impaired in the dysvascular patient. A valid objection to the common use of this procedure in vascular disease is that a short BKA usually can be performed if sufficient soft tissue is available to cover a knee disarticulation. There is a fine line of distinction between sufficient viable skin for coverage of a short below-the-knee residual limb and sufficient viable skin for coverage of a knee disarticulation. A short below-knee residual limb that is otherwise unimpaired is functionally quite superior to knee disarticulation.
  - Until relatively recently, another major disadvantage to knee disarticulation was the inability to provide the patient with a prosthesis that was functional and cosmetic. Recent advances in prosthetic knee mechanisms and socket design now permit swing and stance phase control, and cosmesis, that equal available above-the-knee prostheses.
- 🕮 **Modified knee disarticulation:** Moderate trimming of the femoral condylar prominences and patellofemoral arthrodesis in the intercondylar notch:
  - *Advantages*: Conical residual limb with a large area for end-bearing; allows standard suction-socket fitting without ischial weight bearing

Transfemoral Amputation/Above-the-Knee Amputation

- This was the most common level of amputation in the past because it was easily accomplished in cases of PAD and could more easily assure satisfactory healing. Today, because of the understanding of the principles of site selections, potentials for rehabilitation, and improved fitting methods for amputation at lower levels, AKAs are done much less frequently.
- Approximately 85% of AKAs are secondary to vascular disease.
- Hip flexion contracture is common in AKA, particularly with shorter residual limbs. Up to 20 degrees of flexion can be accommodated in the socket.
- There is an increased healing rate in patients with BKA with vascular disease; therefore, this is preferred to AKA. AKA is thus only indicated in those cases where we expect to find poor tissue viability in knee disarticulation or BKA. It may also be indicated in large tumors of the tibia where the surgeon needs to go to the next level.
- During AKA, the use of myodesis of the adductor muscles will help to maintain adduction position of the femur in the prosthetic socket.

Hip Disarticulation and Hemipelvectomy Amputations

- **Hip disarticulation** is the surgical removal of the entire lower limb by transection through the hip joint.
- **Hemipelvectomy** is the surgical removal of the entire lower limb plus all or a major portion of the ileum.
- Ablative surgery of this magnitude is indicated most often to eradicate a malignant tumor of the bone or soft tissues above the thigh, hip, or pelvic region.

1. Other indications include extensive trauma or uncontrolled infection, especially gas gangrene. On rare occasions, surgical conversion of congenital limb deformity to hip disarticulation may improve prosthetic fit and function.
2. In spite of large functional deficits, it is still possible to fit prostheses for these levels of amputation.

- Hip disarticulation amputation produces a residual limb with an excellent weight-bearing characteristic. Lack of a femoral shaft acting as a lever arm on the prosthesis results in poor mediolateral (ML) trunk stability and marked gait deviations.
- In addition to all the functional deficits created by hip disarticulation, the hemipelvectomy amputee has also lost the excellent weight bearing of the ischial tuberosity, and the prosthesis must bear weight on the semisolid abdominal viscera. Socket design for this amputation level must, therefore, produce weight forces on the abdominal viscera in a manner that is tolerable to the amputee and not injurious to the viscera. Many hemipelvectomy amputees reject the prosthesis because of the bulk and weight of the prosthesis and the energy expenditure required for ambulation when wearing it.

Postoperative Lower Extremity Amputee Evaluation and Management

- When evaluating a patient after an amputation, the history should include the reason for and date of the amputation, dates of revisions, prior ambulatory status, self-care status, cardiopulmonary status, neurologic status, peripheral vascular status, diabetic control, previous surgical procedures, residual limb pain, phantom sensation, and phantom pain.
- Physical examination should include an evaluation of vision and mental status, peripheral vascular status, surgical incision and drainage, skin lesions, residual limb skin mobility, edema and pitting, induration, tenderness, skin redundancy, graft and graft-donor sites, passive ROM, joint stability, sensation, and strength in all extremities.
- 🕮 The ideal shape for transtibial residual limb is *cylindrical*, whereas the ideal transfemoral residual limb is *conical*.
  - Records should be kept of several circumferential measurements of the residual limb at specific distances from the greater trochanter for a transfemoral amputee or from the medial knee joint line for a transtibial amputee. The residual limb, soft tissue status, and bone length should also be assessed.
- One of the keys to success after an amputation is adequate wound healing. Evaluating and optimizing nutrition, anemia, diabetic control, and antibiotic use maximize wound healing. An open incision or wound should be covered with a Telfa pad under the shrinkage device or prosthesis.
- A chronically draining sinus may be the result of a superficial abscess, a bone spur, or localized osteomyelitis. The opening should be probed to determine its depth, and plain films, MRIs, or bone scans should be obtained to determine bony involvement.
- Other postoperative goals of preprosthetic management include:
  1. Pain control
  2. Preparation of residual limb for prosthetic fitting
  3. Maintaining and optimizing ROM, especially in the remaining proximal joints of the amputated extremity
  4. Independent mobility
  5. Independence in self-care and ADLs
  6. Education about prosthetic fitting and care
  7. Support for adaptations to the changes resulting from the amputation
  8. Muscle strengthening should be emphasized and should focus on:
     - Gluteus medius (hip abductor) and gluteus maximus (hip extensor) muscles
     - Any residual hamstring or quadriceps muscles
     - Upper extremities

POSTOPERATIVE RESIDUAL LIMB MANAGEMENT

- A postoperative plaster or fiberglass rigid dressing prevents edema, protects from trauma, and decreases postoperative pain. Postoperative edema occurs within a few minutes, so immediate replacement of the dressing is necessary. Once they are removed for inspection or suture removal, rigid dressings must be replaced within minutes to prevent recurrence of edema.
  1. 🕮 *The removable rigid dressing (RRD)* for the transtibial amputee consists of a plaster or fiberglass cast suspended by a stocking and supracondylar (SC) cuff, and is adjusted by adding or removing socks to maintain compression. Provides good edema control with the advantage of allowing daily inspection. Immediate rigid dressing has quicker healing time than soft dressing and leads to earlier ambulation (Sumpio et al., 2013).
  2. When a rigid dressing is not being used, elastic bandages in a figure-8 wrap should be used. Elastic bandages are less effective shrinkage devices because patients fail to master the wrapping technique, and it requires reapplication many times a day. Poorly applied elastic bandages also can cause circumferential constriction with distal edema. Double-length 4-in. bandages should be used for the transtibial limb and double-length 6-in. bandages for the transfemoral limb.
  3. Elastic shrinker socks are easy to apply and provide uniform compression but are generally used after staples or sutures are removed. They should fit snugly and reach the groin in the transfemoral amputee. They also may cause skin damage with constriction if not properly fitted and maintained.
- The amputee *should wear a shrinkage device 24 hours a day* except for bathing. A shrinkage device for the nonprosthesis candidate helps control pain and edema and facilitates healing. The shrinkage device can be discontinued after fitting the definitive prosthesis if the amputee wears the prosthesis regularly. It can be used at night if overnight edema occurs.
- Contractures are easy to prevent but difficult to correct:
  - *Hip flexion contractures:*
    - The amputee must not lie on an overly soft mattress, use a pillow under the back or thigh, or have the head of the bed elevated. Standing with the transfemoral residual limb resting on a crutch should be avoided. All these practices can lead to hip flexion contractures.
    - Amputees should lie prone for 15 minutes three times a day to prevent hip flexion contractures. The amputee who cannot lie prone should lie supine and actively extend the residual limb while flexing the contralateral leg.
  - *Hip abduction contractures:* The amputee must not place a pillow between the legs, as this can create a hip abduction contracture.
  - *Knee flexion contractures:*
    - A transtibial amputee must not lie with the residual limb hanging over the edge of the bed, with a pillow placed under the knee, or with the knees flexed, and must not sit for a long period of time in order to avoid knee flexion contractures.
    - The transtibial amputee should sit with the knee extended on a board under the wheelchair cushion with a towel wrapped over the board.
- Crutch walking with or without prosthesis promotes good ROM and, when feasible, is preferred over wheelchair mobility.
- After suture removal, the residual limb should be cleansed daily with bland soap and warm water. The limb should be patted completely dry before the application of any shrinkage device.
  - Gentle massage decreases sensitivity to pressure, and deep friction massage perpendicular to the scar prevents scar adhesions.
  - Ensure that the residual limb scar has mobility in all directions. Use a thin layer of emollient to decrease the friction from massage, but discourage the use of thick creams. For very dry skin, a thin emollient can be applied in the evening for absorption overnight.
  - Shaving of the residual limb should be discouraged.

LOWER EXTREMITY PROSTHESIS PRESCRIPTION

- In 1995, Medicare established functional levels of ambulation for amputees. A patient's functional level is determined by the physician and dictates the possible choices of prosthetic components to match the functional level. The patient's functional level should match the K level of the components. The component K level ranges from K0 to K4.

Medicare's Functional Levels of Ambulation for Amputees

FUNCTIONAL LEVEL	COMPONENT (K) LEVEL	DEFINITION	PROSTHESIS COMPONENT(S) ALLOWED
0	K0	Nonambulatory (bedbound)	No prosthesis allowed
1	K1	Limited to transfers or limited household ambulator	Manual lock or stance-control knee, SACH, or single-axis foot
2	K2	Unlimited household but limited community ambulator	Pneumatic or polycentric knee, multiaxis foot
3	K3	Unlimited community ambulator	Hydraulic knee, microprocessor knee, energy-storing foot
4	K4	High-energy activities (sports, work)	

SACH, solid ankle cushion heel.

Lower Extremity Prosthetic Components by Amputation Level

TEMPORARY (PREPARATORY) PROSTHESIS

- A preparatory (temporary) prosthesis is usually made prior to a definitive prosthesis.
- Provides prosthetic fitting before the residual limb volume stabilizes
- Helps in shrinking and shaping of the residual limb
- Allows for early prosthetic training (gait and functional training) and fine-tuning of the prosthetic alignment as the amputee's gait progresses
- May be used as a trial when there is uncertainty about a patient's potential success at using a prosthesis
- Usually used for 3 to 6 months postsurgery—until maximal residual limb shrinkage has been achieved

PERMANENT (DEFINITIVE) PROSTHESIS

- When shaping and shrinking process has ended and residual limb volume has stabilized, a definitive or permanent prosthesis is made.
- A definitive prosthesis will typically need to be replaced about every 3 to 5 years.
- A definitive prosthesis will often have more sophisticated componentry and a cosmetic cover.

PROSTHESIS FOR PARTIAL FOOT AMPUTATIONS

- Small toe amputations do not affect ambulation and usually require no prosthesis.
- Partial foot prostheses are used to restore foot function (particularly walking) and forefoot contour.
- Amputation of the great toe reduces push-off force, thus requiring a stiff sole and toe filler in addition to a molded insole with arch support to maintain the alignment of the amputated foot.
- Partial foot amputations involving the forefoot, such as ray resections and TMAs, generally require only shoe fillers or shoe modifications. The shoe modifications required may include a stiff sole, the addition of a spring steel shank extending to the metatarsal heads, a rocker sole, and/or padding of the tongue of the shoe to help hold the hindfoot firmly in the shoe.
- Transtarsal amputations, such as Chopart, Lisfranc, and Boyd amputations, are not the most desirable levels of elective amputation, but they will have better functional results if there is an active balanced dorsiflexion and plantar flexion with normal skin and heel pad present.

The best prosthetic option for a hindfoot amputation is the use of a custom prosthetic foot with a self-suspending split socket, which allows a regular low-quarter shoe to be worn. A posterior leaf spring (PLS) ankle–foot orthosis (AFO) with partial foot prosthesis is another alternative.

🕮 SYME'S AMPUTATION PROSTHESIS

- In the Syme's amputation, preservation of the articular cartilage covered by the heel pad allows direct end-bearing on this residual limb.
- The patient can stand easily and walk on the end of the residual limb without wearing a prosthesis for short household distances.
- Prosthesis (either posterior or medial opening):
  - Both require removal of a portion of the socket wall to get the bulbous residual limb in.
  - *Major disadvantage:* Poor cosmesis
- Newer socket designs that incorporate an expandable air suspension chamber inside the socket or a thin removable expandable inner socket liner provide a more cosmetically acceptable prosthetic design.
  - *Pros:* Thinner, tighter, and stronger prosthetic socket. There is no need to remove a section from the socket wall, maintaining the structural integrity of the prosthesis.
- 🕮 Prosthetic feet available for a Syme's amputation:
  - Syme solid ankle cushion heel (SACH)
  - Syme stationary ankle flexible endoskeleton (SAFE)
  - Energy-storing carbon fiber foot (low profile)

TRANSTIBIAL AMPUTATION/BELOW-THE-KNEE AMPUTATION PROSTHESIS

- The components in a BKA prosthesis include the socket, the suspension, the shank, and the prosthetic foot.

🕮 PATELLAR TENDON BEARING SOCKET

- The standard socket used for the average BKA amputee is the total-contact patellar tendon bearing (PTB) socket.
- PTB socket is a custom-molded thermoplastic or laminated socket that distributes weight through convex buildups (bulges) over pressure-tolerant areas; it provides concavities (relief areas) on pressure-sensitive areas.
- Characterized by a bar in the anterior wall designed to apply pressure on the patellar tendon
- The trim line extends anteriorly to the mid-patellar level, may extend medially and laterally to the femoral condyles, and extends posteriorly to the level of the PTB bar.
- Despite its name, pressure should be equally distributed over pressure-tolerant areas, with more weight bearing in the area of the patellar tendon and the tibial flare and reduced weight bearing over the bony prominences, such as the tibial crest, distal end of the tibia, and head of the fibula (**Figure 6–10**).

🕮 *PRESSURE-TOLERANT AREAS* (FIGURE 6–10A–C)

1. Patellar tendon
2. Pretibial muscles
3. Popliteal fossa—gastroc-soleus muscles (via gastrocnemius depression)
4. Lateral shaft of fibula
5. Medial tibial flare

🕮 *PRESSURE-SENSITIVE AREAS—RELIEF AREAS* (FIGURE 6–10D–F)

- The following are pressure-sensitive areas where the socket should be adjusted to relieve pressure:
  1. Tibial crest, tubercle, and condyles
  2. Fibular head

3. Distal tibia and fibula
4. Hamstring tendons
5. Patella

- The socket is aligned on the shank on slight flexion (about 5 degrees) to enhance loading of the patellar ligament, prevent genu recurvatum, resist the tendency of the residual limb to slide down the socket, and to place quadriceps muscle in a more efficient and mechanically advantageous position, facilitating its contraction.
- A maximum of 25 degrees of flexion is possible to accommodate knee flexion contracture.
- The alignment of the socket also includes a slight lateral tilt to reduce pressure on the fibular head.
- A liner is added to the socket to protect fragile or insensate skin, reduce shear forces, provide a more comfortable socket for tender residual limbs, or accommodate for growth.
- Liners can be made of closed-cell thermal plastic foams, rubber covered with leather, or silicone/urethane gels. Custom gel liners without the suspension pin have been particularly helpful in managing shear problems that can occur with residual limbs covered with split thickness skin grafts, or boney prominences.

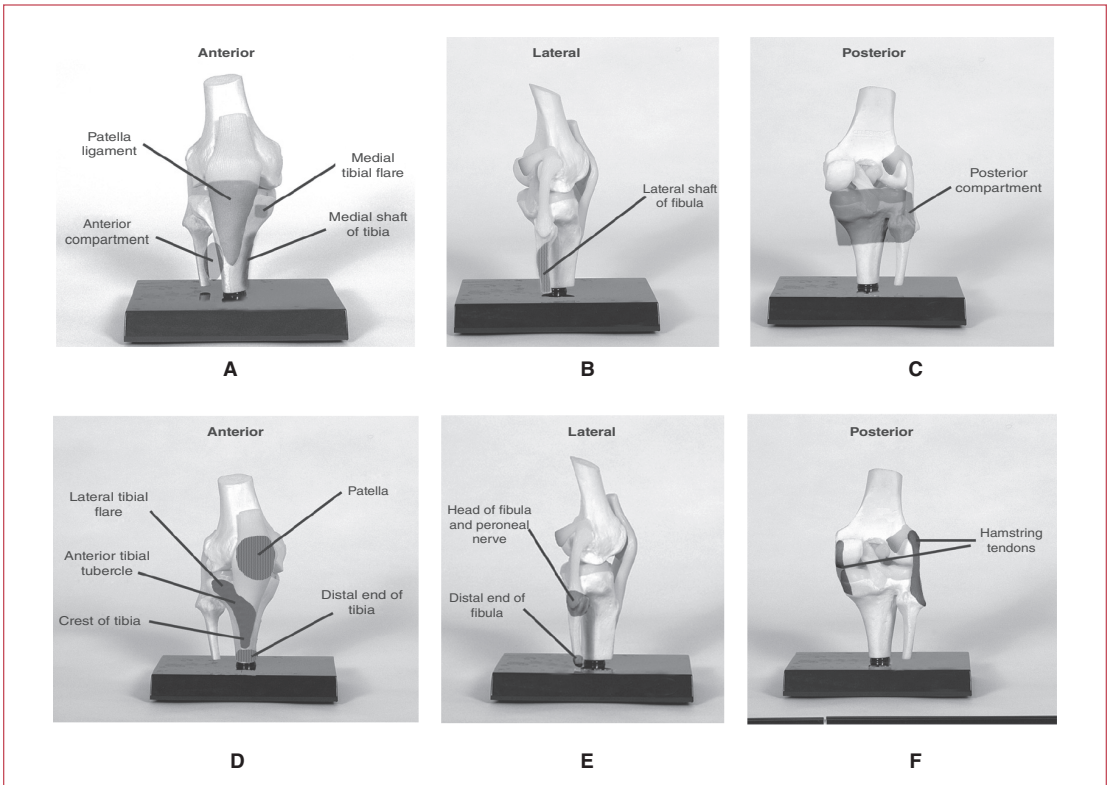


FIGURE 6–10 A, B, and C indicate pressure-tolerant areas in a PTB socket, and D, E, and F indicate pressure relief areas in a PTB socket.
PTB, patellar tendon bearing.

COMMONLY USED SUSPENSION SYSTEMS FOR BELOW-THE-KNEE AMPUTATION

- **Supracondylar cuff suspension socket:**
  - Consists of a cuff or strap that wraps circumferentially around the thigh, fitted immediately above the femoral epicondyles
  - May be used with or without a fork strap and waist-belt suspension
- **Brim suspension:**
  - PTB socket with Supracondylar brim suspension/Supracondylar wedge (PTB-SC):

    - PTB socket with medial and lateral walls extending above the femoral epicondyles (Supracondylar) for suspension
    - Instead of removable medial brim or wedge, a compressible foam wedge buildup may be incorporated into the soft liner.
    - Provides extra ML support and is used for short residual limbs
  - PTB socket with Supracondylar-suprapatellar (PTB-SC/SP) brim suspension:
    - Similar to the PTB-SC socket with the addition of suprapatellar trim lines
    - The suprapatellar trim line improves the suspension.
    - Used for short residual limbs and for controlling genu recurvatum
- **Rubber or neoprene sleeve:**
  - Sleeves made of neoprene, rubber, latex, or other elastic materials may be used as a primary suspension system or in combination with another suspension mechanism.
  - Provides good suspension, fitting snugly over the proximal prosthesis and extending several inches up onto the thigh over the prosthetic sock
  - Should not be used as a primary form of suspension with standard PTB sockets in very short residual limbs, in patients with decreased ML knee stability, or when hyperextension control is required
  - Increased perspiration may be a problem.
- **Pin suspension:**
  - Design includes a silicone or other gel insert or liner with an attached distal pin that locks into the bottom of the plastic laminated socket.
  - Provides excellent suspension for amputees with greater suspension demands, for example, athletes, and for those with short residual limbs, as well as excellent skin protection for the scarred residual limb
  - Expensive because the gel liners are typically replaced annually
- 🕮 **Suction suspension:**
  - Design includes a silicone or other gel insert or liner with the use of a one-way expulsion valve in the distal aspect of the socket. The valve allows air to escape from the socket but not enter.
  - *Advantages:* Does not create pressure distributions that are likely to disturb normal circulation and soft tissue fluid balance as much as pin suspension. Provides excellent suspension for amputees with greater suspension demands, for example, athletes, and for those with short residual limbs, as well as excellent skin protection for the scarred residual limb.
  - Elevated vacuum socket systems use a mechanical or electric pump to generate negative pressure between the limb and the socket for enhanced suspension and contact.
  - *Disadvantage:* Expensive because gel liners are typically replaced annually
- **Thigh corset:**
  - Connects a leather thigh (femoral) corset to a PTB prosthesis through metal joints and side bars to decrease distal residual limb weight bearing by 40% to 60%
  - Used when the patellar tendon cannot tolerate weight bearing or when the knee joint is unstable or painful

PROSTHETIC FEET (TABLE 6–10 AND SEE FIGURE 6–12)

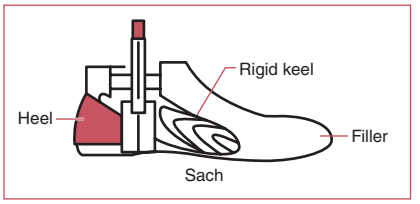
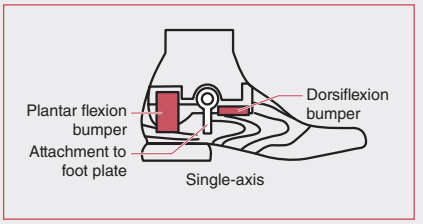
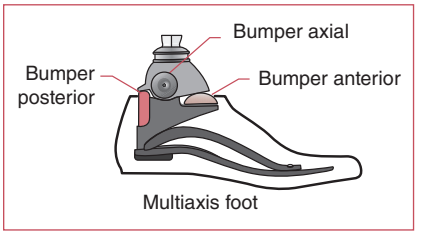
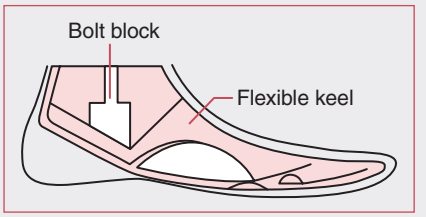
- **SACH foot:**
  - Durable, lightweight, inexpensive, and easily interchanged to accommodate shoes of different heel height
  - 🕮 Compressible heel and wooden keel in this design allow it to simulate the motions of the ankle in normal walking (plantar flexion at heel strike) without actual ankle movement occurring.
  - Accommodates partially for uneven terrain, but this foot is best suited for flat, level surfaces

- **Single-axis foot:**
  - Permits movement of the foot–ankle complex in one plane
  - Movement occurs in the plantar flexion/dorsiflexion axis. The movement is controlled by the use of adjustable internal rubber bumpers that provide resistance to dorsiflexion and plantar flexion.
  - This foot is heavier than the SACH foot, and the internal components need periodic adjustment or replacement.
  - Typically used for more proximal amputations that require additional knee control or stability; can relieve pressure on distal tibia for BKA
- **Multiaxis foot:**
  - Ideal for the amputee who is involved in activities or who walks on uneven terrain
  - Some controlled movement in the normal anatomic planes of the ankle dorsiflexion, plantar flexion, inversion, eversion, and rotation
  - A number of different foot designs are available to provide this motion.
  - Some of the feet accomplish this movement without the use of mechanically moving parts. They rely on the inherent flexibility of the materials and design of the foot. Other multiaxis feet use mechanical systems.
  - The additional mechanical components necessary for the movement of these feet add to their overall weight and may require frequent maintenance, especially in the very active amputee.
  - For the active amputee, however, the improved balance, coordination, and function provided by these feet outweighs the disadvantages of increased weight and more frequent maintenance.
- **Flexible-keel foot:**
  - Mimics multiaxis movement
  - Less maintenance than mechanical multiaxis foot
- **Energy-storing/dynamic-response foot:**
  - Major advances in LE component design continue to be developed, with regards to dynamic-response carbon fiber and fiberglass prosthetic feet.
  - The design of these feet incorporates the use of resilient, flexible, energy-storing materials. Energy is stored in the foot at the time of heel strike, as the weight of the body compresses or flexes the resilient material within the foot and is returned to the amputee at the time of push-off.
  - Resiliency of these feet makes them particularly suitable for amputees involved in activities that require running and jumping. Many amputees report they believe they are more functional using a prosthesis with a dynamic-response foot.
  - These prosthetic designs make ambulation more efficient and require less energy consumption due to their lightweight and energy-storing features.

ANKLE UNITS

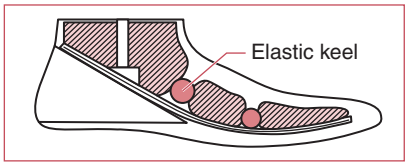
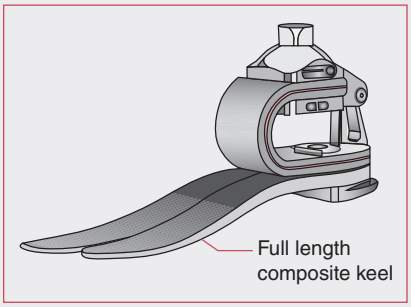
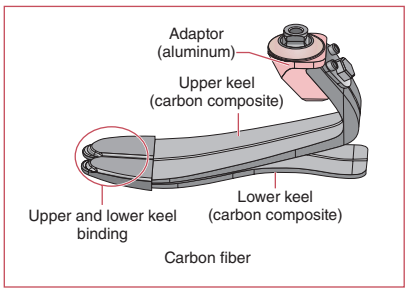
- Multiaxial ankle units, shock absorbers, and torque absorbers can be added to some prosthetic feet.
- Ankle units require increased maintenance and add weight to the prosthesis.
- Hydraulic ankle unit:
  - Cadence-responsive hydraulic ankle unit attached to energy-storing foot allows for walking on uneven terrain, improves ankle ROM for normalization of gait, indicated for K2 and K3 ambulators (Bai et al., 2018).
    - Disadvantages: Cannot be used for running or jumping, increased weight, high cost
- Microprocessor-control hydraulic ankle unit:
  - Microprocessor incorporates sensors and gyroscope to continuously monitor gait cycle and limb positioning to then electronically control ankle ROM, normalizes gait, used for smooth negotiation of uneven terrain, indicated for K2 ambulators (McGrath et al., 2018).
    - Disadvantages: Cannot be used for running or jumping, heaviest, very high cost

TABLE 6–10 Prosthetic Feet

FOOT UNIT	MAIN USE	ADVANTAGES	DISADVANTAGES
RIGID KEEL			
• **SACH** (solid ankle cushion heel) • Wooden keel • Compressible heel 	• K1 component/ Level 1 ambulator • General use • Kids-durable • Limited ambulation needs	• Inexpensive • Light (lightest foot) • Durable • Reliable	• Energy-consuming • Rigid • Best on flat surface
SINGLE-AXIS FOOT			
• Movement in one plane (dorsiflexion and plantar flexion) – Heel height-adjustable single-axis feet available 	• K1 component/ Level 1 ambulator • To enhance knee stability • AKA who needs greater knee stability (goes to flat foot quick before knee buckles); knee goes back into extension (gives stability in early stance)	• Adds stability to prosthetic knees • Increased weight (70% heavier than SACH)	• Increased cost • Increased maintenance
MULTIAXIS FOOT			
• Allow PF (plantarflexion), DF (dorsiflexion), inversion, eversion, and rotation 	• K2 component/ Level 2 ambulator • Used for ambulation on uneven surfaces • Absorbs some of the torsional forces created in ambulation	• Multidirectional motion • Permits some rotation • Accommodates uneven surfaces • Relieves stress on skin and prosthesis	• Relatively bulky • Heavy • Expensive • Increased maintenance • Greater latitude of movement may create instability in patients with ↓ coordination
FLEXIBLE KEEL			
• **SAFE** (stationary ankle flexible endoskeleton) 	• K2 component/ Level 2 ambulator • Used for ambulation on uneven surfaces	• Flexible keel • Multidirectional motion • Moisture-and grit-resistant • Accommodates uneven surfaces • Absorbs rotary torques • Smooth rollover	• Heavy • Increased cost • Not cosmetic • Does not offer inversion/ eversion

(*continued*)

TABLE 6–10 Prosthetic Feet (*continued*)

FOOT UNIT	MAIN USE	ADVANTAGES	DISADVANTAGES
• **STEN** (stored energy)  	• K2 component/ Level 2 ambulator • Used when smooth rollover needed	• Elastic keel • Moderate cost • Accommodates numerous shoe styles • ML stability similar to SACH	• Moderate-heavy weight • Cannot be used with Syme's amputation
ENERGY-STORING FOOT/DYNAMIC RESPONSE			
• **Seattle foot** – Consists of a cantilevered plastic C- or U-shaped keel, which acts as a compressed spring  	• K3 and K4 components/ Level 3 and 4 ambulators • Jogging, general sports, conserves energy	• Energy-storing • Smooth rollover	• High cost • No SACH heel makes it difficult to change compressibility of heel
• **Carbon fiber or fiberglass foot** – Pylon and foot incorporated into one unit – The dynamic keel extends to the bottom of the transtibial socket (and in AKA, to the level of the knee unit)  	• K3 and K4 components/ Level 3 and 4 ambulators • Running, jumping, vigorous sports, conserves energy	• Very light • Most energy-storing • Most stable mediolaterally • Lowest inertia	• Very high cost • Alignment can be cumbersome

AKA, above-the-knee amputation; ML, mediolateral

KNEE DISARTICULATION

- The socket in a knee disarticulation is usually a modified socket with partial end-bearing and some ischial weight bearing, and a soft socket liner with SC buildups to provide suspension.
- The problem with prosthetic fitting of a knee disarticulation is that the prosthetic knee's center of rotation needs to go through the distal residual limb. Fitting a knee unit distal to the residual limb has caused problems in the past, but the four-bar polycentric knee has helped to solve the problem.

- 🕮 The polycentric knee, unlike the single-axis knee, has an instantaneous center of rotation that changes and is proximal and posterior to the knee unit itself. This allows greater knee stability, a more symmetrical gait, and equal knee length when sitting.

TRANSFEMORAL AMPUTATION/ABOVE-THE-KNEE AMPUTATION

- Prostheses for AKA amputees consist of a socket, suspension, knee unit, shank, and foot unit.
- **Sockets for AKA amputees**:
  - The initial design developed specifically for the AKA socket was the total-contact quadrilateral socket. This design replaced the plug fit socket as the standard for above-the-knee prosthesis. Until the mid-1970s, the *quadrilateral socket* was the only socket design used for above-the-knee prosthesis.
  - Now, a more recent socket design is being used. This is the *ischial containment socket* with a narrow ML configuration, and the above the-knee frame socket with a flexible liner known as the ISNY.

🕮 ISCHIAL CONTAINMENT SOCKET (FIGURE 6–11C, D)

- Also known as a narrow ML socket or contoured adducted trochanteric-controlled alignment method (CAT-CAM) socket
- Developed as the normal shape, normal alignment (NSNA) socket by Long; also developed as the CAT-CAM by Sabolich
- ML dimension is narrower than the anterior-posterior measurement.
- Narrow ML design was developed to provide a more normal anatomic (adducted) alignment of the femur inside the socket.
- The ischial tuberosity is contained inside the socket, providing a bony lock between the ischium and greater trochanters.
- Weight bearing is concentrated in the medial aspect of the ischium and the ischial ramus.
- The socket is preflexed 5 to 7 degrees to maximize hip extensor muscle control. A maximum of 20-degree flexion is allowed to accommodate flexion contracture.
- Typically fabricated with a flexible inner thermoplastic liner and a rigid outer frame. Cutouts can be made in the outer frame to accommodate bony areas or sensitive areas.
- *Advantages:*
  - It stabilizes the relationship between the pelvis and proximal femur since the ischial tuberosity is controlled inside the socket rather than on top of the posterior brim like in the quad socket.
  - Narrow ML design is intended to keep the femur in adduction; keeping the femur in adduction during the stance phase permits the hip abductors to be in a more stretched and efficient position.
  - More energy-efficient ambulation at high speed
  - ↑ comfort in groin area compared with quad socket (in which weight bearing is done primarily on the ischial tuberosity)
  - There is less lateral thrust of prosthesis at midstance.
  - Higher trim line results in better control of the residual limb especially for short AKA amputation.
  - Able to accommodate smaller residual limb
  - There is greater medial-lateral control of the pelvis with this design.
- *Disadvantages:*
  - More expensive and difficult to fabricate than the quad socket
  - Wider anterior to posterior dimension at the level of ischial ramus leads to increased movement in anterior/posterior plane.

QUADRILATERAL TRANSFEMORAL SOCKET (QUAD SOCKET; FIGURE 6–11A, B)

- Narrow anterior-posterior and relatively wide medial-lateral dimensions
- Posterior socket has a flat, horizontal shelf on which the ischial tuberosity and the gluteal muscles rest.

- It has a prominent bulge over the Scarpa's triangle for wide pressure distribution.
- There are reliefs for the adductor longus, hamstring, greater trochanter, gluteus maximus, and rectus femoris.
- Ischial tuberosity sits on top of the posterior brim of the quadrilateral transfemoral socket (QUAD) socket.
- Use with hip joint and pelvic band because of lack of stability, especially in geriatric patients
- Low trim line
- *Disadvantages:*
  - ↑ discomfort while seated from either the anterior proximal brim or from the rigid posterior socket wall
  - ↑ skin irritation at ischium and pubis from the proximal brim
  - Tenderness on anterior distal femur
  - ↓ stability compared with narrow ML socket
  - More lateral lurch when walking
  - Poor cosmesis with gapping of lateral socket wall during stance
  - Poor control of the residual femur, allowing lateral shift of the socket

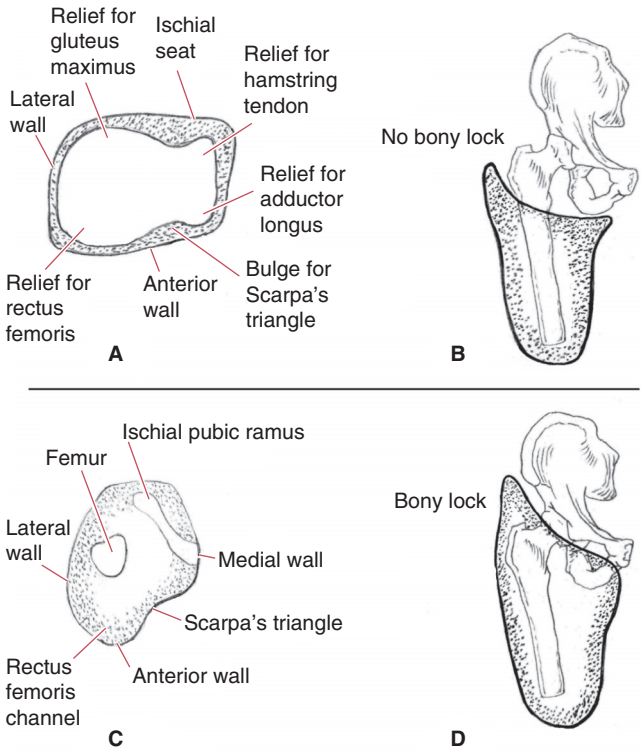


FIGURE 6–11 Transfemoral amputation/above-the-knee amputation socket design. (A, B) Quadrilateral transfemoral socket. (C, D) Ischial containment transfemoral socket.

SOCKET DESIGN FOR TRANSTIBIAL AMPUTATIONS AND TRANSFEMORAL AMPUTATIONS

FLEXIBLE SOCKET

- Provides softer, thermoplastic material for weight transmission
- Sits in a rigid laminated frame
- *Advantages:*
  - Comfort

  - Better sense of outer objects through socket
  - Often translucent or transparent
  - Feels cooler
  - Rapidly fabricated and modified
  - Suction suspension if desired
- *Disadvantages:*
  - Expensive
  - Difficult fit secondary to linear shrinkage of thermoplastic material

GEL LINER/SUCTION SUSPENSION

- Properties similar to human adipose tissue
- *Advantages:*
  - Able to distribute shear forces generated at socket interface
  - Assists vascular inflow by creating a negative pressure
  - Can provide suction suspension
- *Disadvantages:*
  - Not always tolerated, excessive sweating
  - Need constant volume to ensure suction fit
  - Expensive
- There are a variety of prosthetic options for the transfemoral amputee as illustrated in **Figure 6–12.**

SUSPENSION OF ABOVE-THE-KNEE AMPUTATION PROSTHESIS (SEE FIGURE 6–12)

- **Suction socket:**
  - Suction suspension sockets are provided with a one-way valve at the distal end of the socket wall that allows air to escape but not to enter to ensure proper fitting.
  - A pull sock is used over the residual limb; the sock is passed through the valve hole and is used to assist in pulling the limb into the socket.
  - Donning a total suction prosthesis is performed while standing.
  - Suction socket use requires stable residual limb volume (size).
  - If volume ↑, it will prevent fit.
  - If volume ↓, result is loss of suction.
  - **Total suction socket:** Worn without socks on residual limb. It provides the best suspension biomechanically but requires minimal volume fluctuation, good hand strength and dexterity, good balance, and good skin integrity.
  - **Partial suction socket:** Uses a socket with a suction valve, but it is worn with socks, which reduces the airtight suction fit.
  - Provides less suspension than the total suction socket, requiring auxiliary suspension (e.g., Silesian belt or total elastic suspension [TES] belt)
  - Elevated vacuum suspension sockets with a seal-in liner and mechanical or electric pump can provide enhanced suspension, but relies on consistent volume of the residual limb.
- **Gel liners with a pin or strap:** Provide suction to the skin and mechanical attachment to the socket
- **Silesian belt:**
  - Can be worn as the primary form of suspension or as a supplement if suction is not adequate
  - Standard Silesian belt attaches to the posterolateral area of the socket wall (near the greater trochanter) and passes around the back and over the opposite iliac crest where it achieves more of its suspension. It then attaches to the proximal anterior wall at the midline.
  - Provides good control of rotation
- **TES belt:**
  - The TES belt is a neoprene belt attached to the prosthesis and pulled around the waist.
  - Comfortable; enhances the rotational control of the prosthesis
- **Pelvic band and belt with hip joint:**
  - The pelvic band is closely contoured about the anterior iliac crest on the side of the amputation to discourage rotation of the prosthesis on the residual limb during ambulation.
  - Belt use over bypass graft surgery sites and in pregnant women is contraindicated.

KNEE UNITS (FIGURE 6–12; TABLE 6–11)

- All knee units, except for the hydraulic stance-control units, attempt to keep the knee flexion and extension fixed at one angle throughout the stance phase.
- They provide stability during walking, particularly during early knee stance, to prevent the knee from buckling, and allow the knee to bend when the patient sits and, in most cases, during the swing phase (to allow easy clearance of the toes).
- Most are single axis; some are multiaxis.
- **Manual locking knee:**
  - Maximum stability for the debilitated or elderly amputee
  - Worst gait efficiency and increased energy consumption
- **Conventional, single-axis knee:**
  - Light, durable, and inexpensive
  - Rely on alignment for stability and work best at one speed (fixed cadence)
  - Excessive heel rise in the early swing phase, as well as terminal swing impact in the late swing phase, may occur at faster cadences.
  - Amputee must prevent knee buckling by activating hip extensors and maintaining knee in full extension.
  - The debilitated amputee or amputee with a short residual limb cannot adequately contract the hip extensors and requires a knee that is set posterior to the trochanter knee ankle line. This alignment has a disadvantage of causing difficulty in flexion of the knee for the swing phase that causes increased energy expenditure compared with other units.
- **Stance-control knee:**
  - Weight-activated stance-controlled knee may provide a stable stance for up to 20 degrees of flexion by utilizing a friction mechanism within the joint.
  - Design is for amputees with weak hip extensors or for geriatric amputees.
  - Stance control is not automatic, and the amputee must be able to initiate and maintain control of the knee. The knee will buckle if allowed to flex beyond 25 degrees in stance.
- **Four-bar polycentric knee:**
  - Ideal for patients with very long residual limbs as well as for those with poor stability due to short residual limbs, poor balance, or weak hip extensors
  - Fluid control can be added, and some polycentric knees can be manually locked.
- **Hydraulic (fluid controlled) and pneumatic control knee:**
  - Allow for variable cadence and stance phase flexion in gait
  - Pneumatic units are air-filled and are lighter in weight, but they provide only swing phase control.
  - Hydraulic/fluid-controlled knees (oil) are helpful for the active amputee who varies cadence and who can tolerate the extra weight and expense.
  - Fluid-controlled knees can provide swing or swing and stance phase control.
  - When weight is applied to the slightly flexed knee, the unit allows a slow yielding action instead of a precipitous collapse. This permits the young, agile amputee to descend ramps and stairs in a step-over-step manner.
- 🕮 **Microprocessor-control hydraulic knee:**
  - Function is similar to other hydraulic swing and stance phase knees, but it has the additional features of computer-programmed custom settings for each individual.
  - The microprocessor recalibrates the stability of the knee 50 times per second to adapt to changing conditions and ultimately prevent falls.
  - *Advantages:* Computer-adjusting knee for variable gait cycles; energy-saving
  - *Disadvantages:* Highest cost, heavy, increased maintenance, inconvenience of daily charging, and unproven track record for dependability

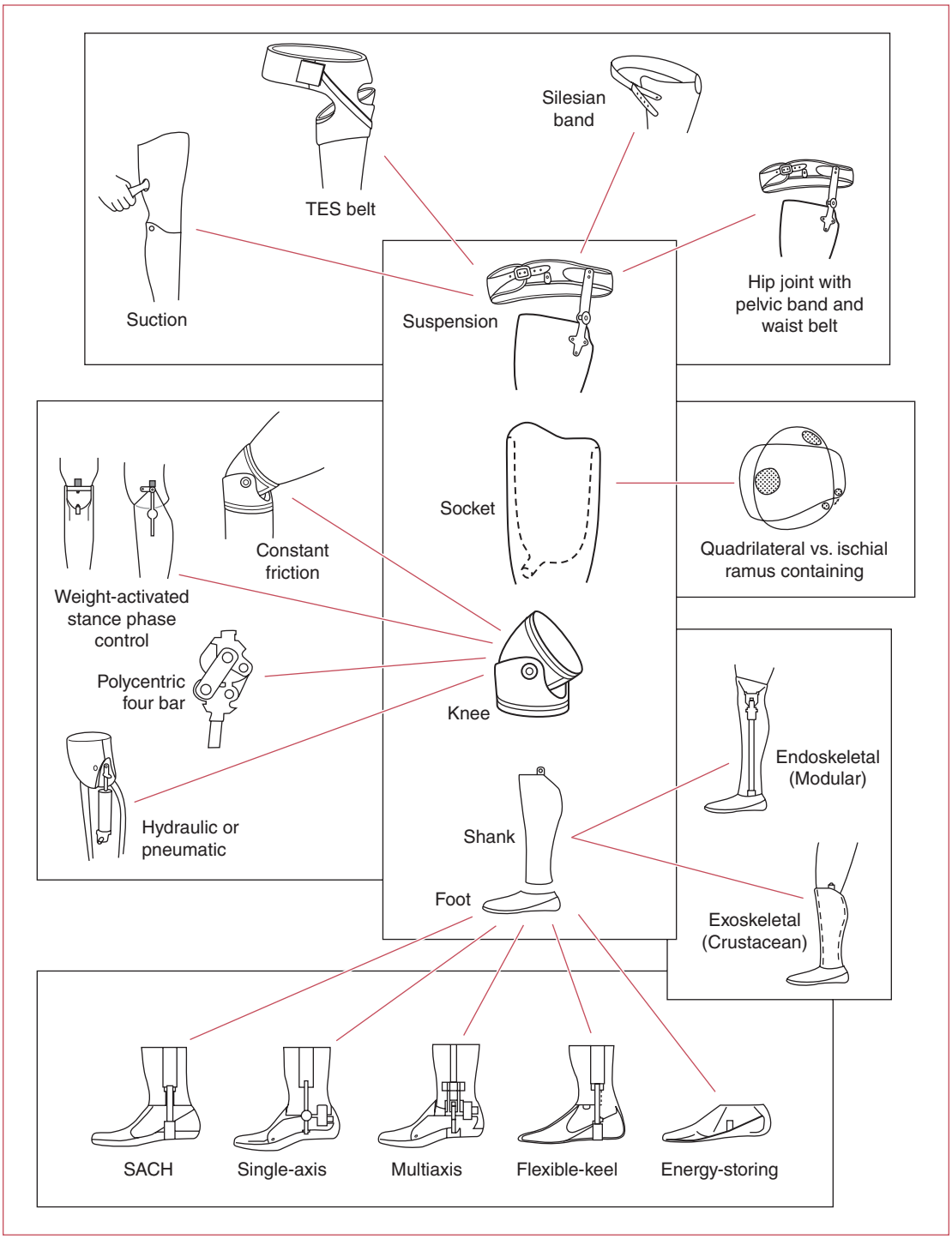


FIGURE 6–12 Transfemoral amputee prosthetic options.
SACH, solid ankle cushion heel; TES, total elastic suspension.
Source: Courtesy of University of Texas Health Science Center at San Antonio.

TABLE 6–11 Knee Units

KNEE UNITS	ADVANTAGES	DISADVANTAGES
CONSTANT FRICTION KNEE (SINGLE AXIS WITH CONSTANT FRICTION UNIT)		
• K1 and K2 components/Level 1 and 2 ambulators • Friction mechanisms are devices used in swing control knee to dampen the pendular action of the prosthetic knee during the swing phase, to decrease the incidence of high heel rise in early swing, and decrease terminal impact in late swing • Single walking speed • May be used in kids • No stance control; a screw used to adjust the friction to determine how fast or slow the knee swings	• Inexpensive • Durable • Reliable	• Low stability (in early stance, the single axis has the lowest zone of stability) • Fixed single cadence • Too much friction prevents knee from flexing • Too little friction causes the knee to swing too easily; patient needs to vault • No stance control
STANCE-CONTROL KNEE/SAFETY KNEE/WEIGHT-ACTIVATED FRICTION BRAKE		
• K1 and K2 components/Level 1 and 2 ambulators • Single-axis knee with stance control • Stance control acts as a brake system • Indications: – Geriatrics – Short residual limb – General disability – Uneven surfaces – Amputees with weak hip extensors	• Improved knee stability • Braking mechanism if weight applied with knee flexed 0–20 degrees	• Slightly increased weight • Increased cost • Increased maintenance • Must unload fully to flex • Cannot use in bilateral AKA (knees would not bend with loading) → cannot bend both knees at the same time (patient cannot sit down) • Activities that require knee motion under weight bearing, such as step-over-step stair descent, are incompatible with this knee
POLYCENTRIC/FOUR-BAR KNEE		
• K1 and K2 components/Level 1 and 2 ambulators; no stance control, but inherently stable • Short knee unit → can be used in knee disarticulation and long residual limb	• Excellent knee stability • Improved cosmesis in knee disarticulation and long residual limb patients	• Greater weight, bulk, cost, maintenance • Although durable, needs maintenance every 3–6 months
MANUAL LOCKING KNEE		
• K1 and K2 components/Level 1 and 2 ambulators. • Knee of last resort used in: 1. Blind 2. Stroke patient with amputation • Commonly uses a spring-loaded pin that automatically locks the knee when the amputee stands or extends the knee • Knee is kept extended throughout the entire gait cycle to ↑ stability	• Ultimate knee stability in stance phase	• Abnormal gait • Awkward sitting • Cannot use in bilateral AKA because, like the stance-control knee, patients cannot sit. Knees lock on loading; therefore, the individual cannot bend both knees at the same time to sit down.

(*continued*)

TABLE 6–11 Knee Units (*continued*)

🕮 FLUID-CONTROLLED KNEE UNITS		
• K3 and K4 components/Level 3 and 4 ambulators 1. Hydraulic (oil) 2. Pneumatic (air) – Cadence-responsive knee units through cadence-dependent resistance – 🕮 Allows for either swing phase, or swing and stance phase control – Indications: a. For patients who vary cadence frequency b. Active walkers c. Ambulation in uneven terrain	• Variable cadence • Smoothest gait • Stable; will not lock unless at full extension • Pneumatic units are lighter but hydraulic units tolerate more weight (can support the heavier and more athletic amputee) • Can unlock for some activities (e.g., biking)	• Greatest weight • Increased cost • Increased maintenance
🕮 MICROPROCESSOR-CONTROL HYDRAULIC KNEE		
• K3 and K4 components/Level 3 and 4 ambulators • Function similar to other hydraulic swing and stance phase knees • Has additional features of computer-programmed custom settings for each individual • Microprocessor recalibrates the stability of the knee 50 times per second to adapt to changing conditions and ultimately prevent falls	• Computer-adjusting knee for variable gait cycles • Energy-saving	• Highest cost • Heavy • Increased maintenance • Inconvenience of daily charging • Unproven track record for dependability

AKA, above-the-knee amputation.

HIP DISARTICULATION/HEMIPELVECTOMY

- Amputees with <5 cm of residual femur usually are fitted as hip disarticulation level amputees.
- A **Canadian hip disarticulation prosthesis** is the standard prosthesis for a hip disarticulation.
  - The socket of this prosthesis encloses the hemipelvis on the side of the amputation and extends around the hemipelvis of the nonamputated side, leaving an opening for the nonamputated LE.
  - There is a flexible anterior wall with an opening that allows the prosthesis to be donned. Weight bearing is on the ischial tuberosity of the amputated side.
  - Endoskeletal prosthetic components are preferred for this level of amputation to reduce the overall weight. The endoskeletal hip joint has an extension assist as does the knee unit, which usually is a stance-control knee.
  - Endoskeletal components may be made from aluminum, titanium, or carbon graphite composite.
  - Single-axis foot or SACH foot with a soft heel is used most often. The newer lightweight foot–ankle combination may be even a better option.
- The prosthesis for a hemipelvectomy resembles that for the hip disarticulation socket except in the interior configuration of the socket.
  - In the hemipelvectomy, most of the weight bearing is by the soft tissues of the amputated side, with some of the weight bearing by the sacrum on the opposite ischial tuberosity.

🕮 SWIM PROSTHESES

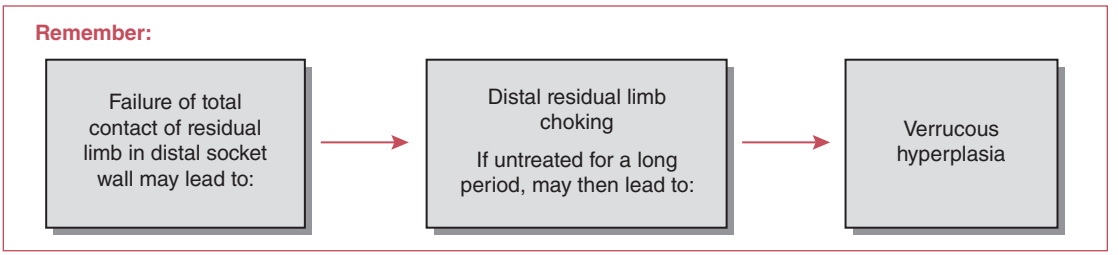
- Waterproof prosthesis or special swim leg
- Peg leg with or without fin attached for BKA or AKA

- Rubber suspension sleeve used for BKA (ActiveSleeve, Michigan Sleeve, Ottobock Swimming Sleeve) prevents water from entering the socket.
- Hollow-chambered leg for BKA or AKA
- Exoskeletal leg with knee lock for AKA
- Waterproof suspension belt for AKA
- Flat foot for barefoot walking for BKA or AKA (Kingsley Beachcomber foot)

COMMON LOWER EXTREMITY AMPUTEE PROBLEMS AND COMPLICATIONS

Dermatologic Issues

- Numerous skin problems can occur on the amputee's residual limb, including folliculitis, allergic dermatitis, hyperhidrosis, and fungal infections. Skin lesions of the residual limb can expand rapidly, so early intervention is required, particularly for patients with diabetes.
- Careful daily inspection of the residual limb should be done, as well as daily residual limb and socket washing.
  1. *Folliculitis* is a common problem in the amputee. It is a hair root infection resulting from poor hygiene, sweating, poor socket fit, or pistoning. It is important to clean the area with antiseptic cleanser, to keep it dry, and to consider administration of oral antibiotics.
  2. *Boils* and *abscesses* should be treated with limited prosthetic use, incision and drainage, and oral antibiotics.
  3. *Epidermoid cysts* occur when sebaceous glands are plugged by keratin and usually are not apparent until months or years after a prosthesis is worn. They grow up to 5 cm in diameter and may break to discharge purulent fluid. They may require incision and drainage.
  4. *Tinea corporis* and *tinea cruris* mainly result from sweating; they may be confirmed through culture or microscopy and are treated by topical or oral fungicides, as well as by good residual limb and socket hygiene.
  5. *Hyperhidrosis* (excessive sweating) of the residual limb is a common problem after amputation. Increased sweating on the residual limb may cause skin maceration, which in turn predisposes the skin to infection by bacteria and fungi and injury by outside forces. Hyperhidrosis can be controlled with antiperspirants (e.g., Drysol or Certain Dri); astringents and rubbing alcohol should be avoided as they dry the skin excessively.
  6. *Allergic dermatitis* may arise from the detergents used to wash the limb socks, from lotions and topical medications, or from agents used in the prosthetic manufacturing process. The allergic dermatitis should resolve with cessation of contact with the offending agent. Eczema may appear acutely with small blisters and later with scaling and erythema. Topical corticosteroids should be applied and the offending agent should be identified and removed.
  7. 🕮 *Choke syndrome* is caused by the proximal prosthetic socket being too tight and lack of total contact between residual limb and the socket leads to impairment of venous return. It occurs most commonly at the distal transtibial residual limb due to atrophy over time and use of additional socks or when the residual limb becomes larger (excessive weight gain).
     - Findings on exam: Initially with wearing a prosthesis, the patient develops a well-circumscribed indurated area; if acute, edema can have weeping/blistering of the skin. Area is tender to palpation and prone to cellulitis. When chronic, the skin becomes thickened and hyperpigmented due to hemosiderin accumulation.
     - Treatment: Relieving the proximal constriction and restoring total contact between the socket and residual limb by reducing the number of socks and modification of the distal end pad. Often a new total contact socket is needed.
  8. 🕮 *Verrucous hyperplasia* is a wartlike skin overgrowth, usually of the residual distal limb, resulting from inadequate socket wall contact with subsequent edema formation. A chronic *choke syndrome* may lead to verrucous hyperplasia of the distal residual limb skin.
     - Relieving the proximal constriction and reestablishing total contact within the socket can reverse these processes.



Bone Problems

- If the periosteum is incorrectly stripped during surgery or if it was stripped during trauma, bone *spurs* causing pressure on the skin and pain may arise. Socket modifications generally take care of the problem. However, in some cases, additional surgery may be needed.
- Bone pain may also result from a hypermobile fibula that is left longer than the tibia. If a balanced myodesis was not performed in a transfemoral amputation (TFA), the femur may extrude through the muscle and present subcutaneously. If prosthetic adjustments, such as a flexible socket, are inadequate for the extruded femur, then surgical intervention might be needed.
- Bone overgrowth and heterotopic ossification occur most commonly in kids with acquired amputation and young adults with traumatic amputation.

Pain in the Amputee Patient

- Pain in the amputee patient can be broken down into incisional pain on the residual limb, phantom limb sensation (*nonpainful*), and phantom pain.

1. INCISIONAL PAIN

- Incisional pain should subside with healing, although shear forces on adherent scars or bone spurs can be painful. Deep massage helps prevent scar adhesions. Local pain may be due to an unprotected neuroma (nerve ending left "exposed" during surgery) that is being pressed upon. Neuroma exposed to pressure may give great pain and may require revision if unresponsive to conservative treatment.
- Remember that not all residual limb pain is a result of a problem within the amputation site or due to a poorly fitting prosthesis.
  - The amputee may experience intermittent claudication pain in parts of the residual limb. Patients with herniated discs may feel referred pain in a specific segmental distribution even after those body parts have been removed.
  - After *tumor* amputation, local recurrence of the tumor might also be an explanation for a painful residual limb.

2. PHANTOM LIMB SENSATION

- All people with an acquired amputation will experience some form of phantom limb sensation. It is a normal occurrence after amputation of a limb. Phantom sensation is an awareness of a *nonpainful* sensation in the amputated part (distal to the site of amputation). Phantom sensation usually diminishes with time but can persist throughout the amputee's life. No treatment is required.

3. PHANTOM LIMB PAIN

- This involves awareness of pain or any noxious stimulus in the portion of the extremity that has been amputated. It may accompany the phantom sensation, localizing in the phantom limb rather than in the residual limb.
- The pain has been described as cramping, aching, burning, and occasionally lancinating.
- Etiology: Phantom pain appears to be related to neuron *deafferentation hyperexcitability*.
- It may be diffuse throughout the entire limb or may be localized to a single-nerve distribution.
- Studies have suggested that 50% to 85% of amputees experience some phantom limb pain.
- Recent data do not suggest a predisposition for phantom limb pain among traumatic amputees, older adult amputees, or those with pain in the amputated limb before amputation.

There appears to be no correlation between phantom pain and amount of time after amputation or use of prosthesis.
- Phantom pain usually diminishes with time, and chronic phantom pain is rare. The occurrence of phantom pain generally is considered to be a significant long-term problem in only 5% or less of the total amputee population.
- If pain persists >6 months, prognosis for spontaneous recovery is poor.
- Phantom pain does not occur in congenital limb deficiency.

TREATMENT

- Physical interventions/physical modalities usually offer temporary relief. Relief is probably mediated by the gate control mechanism:
  - Acupuncture
  - Transcutaneous electrical nerve stimulation (TENS)
  - Vibration
  - Ultrasound
- Medical intervention (including neuropharmacologic intervention):
  - Acetaminophen and nonsteroidal anti-inflammatory drugs (NSAIDs): A cross-sectional study found acetaminophen and NSAIDs to be the most common medications used in phantom limb pain (Subedi & Grossberg, 2011).
  - Tricyclic antidepressants
  - Gamma-aminobutyric acid (GABA) inhibitors
  - Serotonin-selective reuptake inhibitors/serotonin and norepinephrine reuptake inhibitors (SSRIs/SNRIs)
  - Anticonvulsants
  - Calcitonin
  - Capsaicin
  - Propanolol
  - Mexiletine—Na^+ channel blocker
- Psychological intervention:
  - Hypnosis
  - Biofeedback
  - Cognitive therapy
  - Behavioral
  - Support groups
  - Relaxation therapy
  - Voluntary control of the phantom limb (mental imaging)
- Procedures and surgical intervention:
  - Usually less favorable, with poor long-term success
  - Severe cases may need nerve blocks, steroid injections, or epidural blocks.
  - Regional guanethidine or reserpine blocks have been tried with little success.
  - Sympathectomy and other neurosurgical procedures

Common Causes of Prosthetic Gait Deviations by Amputation Level

1. **Partial foot amputee:** The average patient with a partial foot amputation will require little gait training unless other parts of the body are involved.
2. **Syme's amputee:** The patient has usually walked in a plaster cast with a rubber heel or artificial foot prior to delivery of the definitive prosthesis. Minimal training is required during the cast period, except for occasional use of crutches or a pick-up walker at the outset.
   - Gait speed is typically decreased 32%.
   - Oxygen consumption increased 13% per distance walked in the vascular Syme's amputee with prosthesis, compared with normal subjects without vascular disease.
3. 🕮 **Transtibial amputee gait deviations (Tables 6–12 and 6–13):**
   - Excessive knee flexion
   - Excessive knee extension

TABLE 6–12 Transtibial Amputee Gait Deviations: Excessive Flexion/Extension Movements

EXCESSIVE KNEE FLEXION (↑ KNEE FLEXION AT MOMENT OF INITIAL CONTACT)	EXCESSIVE KNEE EXTENSION (RECURVATUM; ↑ KNEE EXTENSION AT MOMENT OF INITIAL CONTACT)
• ↑ Ankle dorsiflexion	↑ Ankle plantar flexion
• Excessive anterior displacement of the socket over the foot • Moving *socket* anteriorly in relation to the foot	Moving *socket* posteriorly in relation to the foot
• Excessive *posterior* displacement of the foot in relation to the socket • Moving *foot* posteriorly in relation to the socket	Moving *foot* anteriorly in relation to the socket
• Too hard heel cushion (or plantar flexion bumper)	Too soft heel cushion (or plantar flexion bumper)
• Knee flexion contracture	Quad weakness (excessive knee extension used as a stabilizing technique)
	Distal anterior tibial discomfort
	Habit

TABLE 6–13 Gait Analysis of the Transtibial Amputee

PROBLEM	CAUSE	SOLUTION
Delayed, abrupt, and limited knee flexion after heel strike	Heel wedge is too soft, foot is too far anterior	Stiffen heel wedge, move foot posterior
Extended knee throughout stance phase	Too much plantar flexion	Dorsiflex foot
Toe stays off the floor after initial contact (heel strike)	Heel wedge too stiff, foot too anterior, too much dorsiflexion	Soften heel wedge, move foot posterior, plantar flex foot
"Hill-climbing" sensation toward end of stance phase	Foot too anterior, too much plantar flexion	Move foot posterior, dorsiflex foot
High pressure against patella throughout most of stance phase, heel is off the floor when patient stands	Foot too plantar flexed	Dorsiflex foot
Knee too forcefully and rapidly flexed after heel strike, high pressure against anterodistal tibia at heel strike, and/or prolonged discomfort at this point	Heel wedge too stiff, foot too far posterior, foot too dorsiflexed	Soften heel, move foot anterior, plantar flex foot
Hips level, but prosthesis seems short	Foot too far posterior, foot too dorsiflexed	Move foot anterior, plantar flex foot
Drop-off at end of stance phase	Foot too far posterior	Move foot anterior
Toe off of floor as patient stands or knee flexed too much	Foot too dorsiflexed	Plantar flex foot
Valgus moment at knee (knock-knee) during stance phase, excessive pressure on distal medial limb and proximal lateral surface of knee	Foot too outset	Inset foot
Excessive varus moment at knee (bow-legged) during stance phase (a varus moment at the knee should occur in stance phase but should never be excessive); the distal lateral residual limb is painful	Mediolateral dimension of socket too large, foot too inset	Fit of socket should be checked, outset foot

- *Excessive knee varus moment during stance phase/excessive lateral thrust of the socket brim:*
  - Foot too inset (excessive medial placement of prosthetic foot in relation to socket)
  - Abducted socket
  - Patient may complain of pain/pressure on proximal medial or distal lateral aspect of residual limb.
- *Excessive knee valgus moment during stance phase:*
  - Foot too outset (excessive lateral placement of prosthetic foot in relation to socket)
  - Adducted socket
  - Patient may complain of pain/pressure on proximal-lateral or distal-medial aspect of residual limb.

4. 🕮 **Transfemoral amputee gait deviations (Table 6–14):**
   a. Lateral trunk bending
   b. Abducted gait
   c. Circumduction
   d. Vaulting
   e. Medial and lateral whips (swing phase)
   f. Foot rotation at heel strike
   g. Foot slap
   h. Uneven heel rise
   i. Terminal impact
   j. Uneven step length
   k. Exaggerated lordosis
   l. Instability of prosthetic knee during stance
   m. Drop-off at end of stance phase
5. **Hip disarticulation:**
   - During the course of gait training, various prosthetic gait deviations may become obvious.
   - *Knee instability* in the stance phase (caused by increased knee flexion moment) may be due to improper alignment of the prosthesis so that the weight-bearing line passes posterior to the knee axis of motion, the plantar flexion bumper of the articulated foot or the heel cushion of the SACH being too firm, or the hip bumper contacting the socket too soon.
   - In contrast, difficulty in flexing the knee will occur if the knee axis of motion is placed too far posterior to the weight-bearing line. Excessive knee flexion in the swing phase occurs if the extension aid is too weak or the friction in the knee bolt is inadequate.
   - Circumduction of the prosthesis during swing phase or vaulting on the sound side may be due to excessive length of the prosthesis, inadequate suspension, or excessive knee instability.

Pediatric/Juvenile Amputees

PROSTHETIC MANAGEMENT OF THE JUVENILE AMPUTEE

- *Prosthetic training* for children is different from that for adults. The child's level of development and attention span must be considered when planning training sessions. Prosthetic training frequently must be included as part of play activities. Parents are taught how to assist their child in attaining skills necessary to use the prosthesis successfully. Parent acceptance of the prosthesis is a prerequisite to the child's acceptance.
- *Congenital limb deficiency*: Adults always lose a limb, whereas some children are born with a congenital limb deficiency. In such children, there is no period of psychological adjustment or sense of loss. The prosthesis is perceived as an aid rather than a replacement and will be discarded if it is not helpful.

TABLE 6–14 Gait Analysis of the Transfemoral Amputee

PROBLEM AND CHARACTERISTICS	PROSTHETIC CAUSES	AMPUTEE CAUSES
a. Lateral bending of trunk: Excessive bending occurs laterally from midline, generally to prosthetic side	• Prosthesis may be too short. • Improperly shaped lateral wall may fail to provide adequate support for femur. • High medial wall may cause amputee to lean away to minimize discomfort. • Prosthesis aligned in abduction may cause wide-based gait, resulting in this defect.	• Amputee may not have adequate balance. • Amputee may have hip abduction contracture. • Residual limb may be oversensitive and painful. • Very short residual limb may fail to provide a sufficient lever arm for pelvis. • Defect may be due to habit pattern.
b. Abducted gait: Very wide-based gait with prosthesis held away from midline at all times	• Prosthesis may be too long. • Too much abduction may have been built into prosthesis. • High medial wall may cause amputee to hold prosthesis away to avoid ramus pressure. • Improperly shaped lateral wall can fail to provide adequate support to the femur. • Pelvic band may be positioned too far away from patient's body.	• Patient may have hip abduction contracture. • Defect may be due to habit pattern.
c. Circumducted gait: Prosthesis swings laterally in wide area during swing phase	• Prosthesis may be too long. • Prosthesis may have too much alignment stability or friction in knee, making it difficult to bend knee in swing-through.	• Inadequate suspension. • Amputee may have abduction contracture of residual limb. • Patient may lack confidence for flexing prosthetic knee because of muscle weakness or fear of stubbing toe and falling. • Defect may be due to habit pattern.
d. Vaulting: Rising on toe of sound foot permits amputee to swing prosthesis through with little knee flexion	• Prosthesis may be too long. • Socket suspension may be inadequate. • Excessive stability in alignment or some limitation of knee flexion, such as knee lock or strong extension aid, may cause this deficit.	• Vaulting is a fairly frequent habit pattern. • Fear of stubbing toe may cause this defect. • Residual limb discomfort may be a factor.
e. Medial or lateral whips: Whips best observed when patient walks away from observer; a medial whip is present when heel travels medially on initial flexion at beginning of swing phase; a lateral whip exists when heel moves laterally	• Lateral whips may result from excessive internal rotation of prosthetic knee. • Medial whips may result from excessive external rotation of knee. • Socket may fit too tightly, thus reflecting residual limb rotation. • Excessive valgus or "knock" in prosthetic knee may contribute to this defect. • Badly aligned toe break in a conventional foot may cause twisting on toe-off.	• Improper donning of socket or socket rotated on limb

(*continued*)

TABLE 6–14 Gait Analysis of the Transfemoral Amputee (*continued*)

PROBLEM AND CHARACTERISTICS	PROSTHETIC CAUSES	AMPUTEE CAUSES
f. Foot rotation at heel strike: As heel contacts the ground, the foot rotates laterally, sometimes with vibrating motion	• Too hard heel cushion or plantar flexion bumper	• Weakness at hip muscles
g. Foot slap: The foot plantar flexes too rapidly and strikes the floor with a slap	• Plantar flexion bumper is too soft, offering insufficient resistance to foot motion as weight is transferred to the prosthesis.	• None
h. Uneven heel rise: Prosthetic heel rises quite markedly and rapidly when knee is flexed at beginning of swing phase	• Knee joint may have insufficient friction. • Extension aid may be inadequate.	• Amputee may be using more power than necessary to force knee into flexion.
i. Terminal swing impact: Rapid forward movement of shin piece allows knee to reach maximum force before heel strike	• Knee friction is insufficient. • Knee extension aid may be too strong.	• Amputee may try to assure themselves that knee is in full extension by deliberately and forcibly extending the residual limb.
j. Uneven step length: The length of the step taken with the prosthesis differs from that of the sound leg	• Insufficient socket flexion • Insufficient friction at the prosthetic knee or too loose an extension aid	• Pain or insecurity causing amputee to transfer weight quickly from the prosthesis to the sound leg • Hip flexion contracture
k. Exaggerated lordosis: The lumbar lordosis is exaggerated when the prosthesis is in stance phase, and the trunk may lean posteriorly	• Insufficient socket flexion • Insufficient support from the anterior socket brim	• Hip flexion contracture • Weak hip extensors • Weak abdominal muscles
l. Instability of the prosthetic knee creating a danger of falling	• Knee joint may be too far ahead of trochanter-knee-ankle line. • Socket is mounted with excess flexion. • Plantar flexion resistance may be too great, causing knee to buckle at heel strike. • Failure to limit dorsiflexion can lead to incomplete knee control.	• Amputee may have hip extensor weakness. • Severe hip flexion contracture may cause instability.
m. Drop-off at end of stance phase; downward movement of trunk as body moves forward over prosthesis	• Limitation of dorsiflexion of prosthetic foot is inadequate. • Heel of SACH-type foot may be too short, or toe break of a conventional foot may be too far posterior. • Socket may have been placed too far anterior in relation to foot.	• None

SACH, Syme solid ankle cushion heel.
Source: From Braddom RL. *Physical Medicine and Rehabilitation*. W. B. Saunders; 1996:334–336, 1194–1196, with permission.

- *Prosthetic fitting*: **Table 6–15** gives an overview of the key components for proper pediatric prosthetic fitting. Prostheses fitting should generally occur in the fall, after the summer growth period.

TABLE 6–15 Pediatric Prosthetic Fitting

LEVEL OF PEDIATRIC AMPUTATION	AGE FOR PROSTHETIC FITTING	DEVELOPMENTAL MILESTONES	PROSTHETIC PRESCRIPTION
Transradial	6 months	Child can sit and reaches across midline for bimanual manipulation of objects	Body power—passive mitt terminal device, plastic laminate, self-suspending socket
	9 months		External-power—cookie-crusher, single-site control (one movement flexor)
	18 months		Go to two-site control
Transhumeral	6 months	Same as transradial	Body power—passive mitt and elbow, activate elbow at 18–36 months
	24 months		External-power—variety village elbow with two-site control
Transtibial	9–12 months	Child pulls to stand	PTB, plastic laminate, supracondylar strap, SACH foot, pediatric dynamic responsive feet now available for use
Transfemoral	9–12 months	Child pulls to stand	Narrow mediolaterally, ischial containment with no knee unit, suspension with Silesian bandage, add knee unit at 18 months

PTB, patellar tendon bearing; SACH, solid ankle cushion heel.
Source: From DeLisa JA, Gans BA. *Rehabilitation Medicine: Principles and Practice*. 3rd ed. Lippincott-Raven; 1998:167–187, 635–696, with permission.

- *Prosthetic adjustments/replacements*: Pediatric amputees require frequent prosthetic adjustment and replacements. This is due to normal growth and more rigorous use of the device. It can be expected that a socket or prosthesis will have to be replaced yearly in the first 5 years of life, every 18 months for ages 5 to 12, and every 2 years until the age of 21.
  - To address growth problems, multilayered sockets (onion sockets) for body-powered devices can be used. It allows removal of one layer at a time to accommodate growth. A socket made this way can increase the life span of the prosthesis to 6 to 18 months. Length adjustment is also important.
  - This can be adjusted by adding material to the wrist or elbow sites. Harnesses and cables need to be adjusted for length and replaced more frequently.

🕮 PROSTHETIC REPLACEMENT FREQUENCY IN THE PEDIATRIC AMPUTEE	
First 5 years of age	Yearly
Ages 5–12	Every 18 months
Ages 12–21	Every 2 years

- 🕮 *Bony overgrowth:*
  - Much more common in *acquired amputations in children* than in adults
  - Bone usually grows faster than the overlying skin and soft tissue in the distal end of amputated long bones (residual limb).

  - Formation of bursa may occur over the sharp end and at times the bone may actually protrude through the skin.
  - Overgrowth is seen most frequently in the humerus, fibula, tibia, and femur (in that order).
  - Overgrowth has been reported in congenital limb deficiencies but very rarely.
  - This may require surgical revision several times before skeletal maturity. Different techniques are available, including capping the end of the involved long bone with a cartilage epiphysis to eliminate the overgrowth.
- *Parental counseling and support:* This is key because the acceptance of the prosthesis by the child is often dependent on the acceptance by the parents.

ASSISTIVE DEVICES (AMBULATION AIDS)

CANES

- Require patients have adequate balance, upper body strength, and dexterity for safe use (Sehgal et al., 2021)
- When used properly, canes will:
  - Increase base of support
  - Decrease loading and demand on the lower limbs
  - Provide additional sensory feedback
  - Assist acceleration/deceleration during locomotion
- Prescribed in various disabilities to:
  - Improve balance
  - Increase standing tolerance (Sehgal et al., 2021)
  - Decrease pain
  - Reduce weight-bearing forces on injured structures
  - Compensate for weak muscles
  - Scan the immediate environment
  - 🕮 Offers least stability out of assistive devices (Sehgal et al., 2021)

COMPONENTS

- Handle
- Adjusting knob for handle
- Shaft
- Adjustable mechanism for height
- Rubber tip

C-Handle (Crook Top) Cane

- *Advantages:* Inexpensive
- *Disadvantages:* Uncomfortable, difficult to grasp (e.g., rheumatoid arthritis [RA] patient), weight-bearing line falls behind the shaft of the cane reducing support, not adjustable

Adjustable Metal Cane

- *Advantages:* Inexpensive, adjustable
- *Disadvantages:* Same as earlier

Functional Grip Cane

- *Advantages:* Straight grip handle, improved grip, conforms to natural angle of hand, weight-bearing line is more centered over the shaft of the cane, provides more support
- *Disadvantages:* More expensive

Small-Based Quad Cane (SBQC)

- 🕮 *Advantages:* Fits on stairs, has increased base of support compared with a straight cane, and more effective than straight cane in preventing postural sway in hemiparetic patients during stance (Laufer, 2003).
- *Disadvantages:* Not as stable as a wide-based quad cane (WBQC)

Wide-Based Quad Cane

- *Advantages:* More support, wider base of support
- *Disadvantages:* Heavy, awkward appearance; WBQC does not fit on stairs.

CANE MEASUREMENT/PRESCRIPTION

- 🕮 20- to 30-degree elbow flexion or height of the greater trochanter of the hip for cane height

Difference Between a Crutch and a Cane?

- Cane has one point of contact with the body.
- Crutch has two points of contact with the body.

CRUTCHES

Axillary Crutches

COMPONENTS

- Padded axillary piece (on top)
- Two upright shafts
- Hand piece (in middle)
- Extension piece (adjustable)
- Rubber tip
- *Advantages:* Inexpensive, adjustable, easy to use
- *Disadvantages:* Need good strength and ROM in upper limbs; ties up hands; increased cardiac/metabolic demand; risk of nerve compression in the axilla from improper use

Forearm Crutches (Lofstrand Crutches)

COMPONENTS

- Forearm cuff with narrow anterior opening
- Forearm piece bent posteriorly and adjustable (extends to 2 in. below elbow)
- Molded hand piece
- Single aluminum tubular shaft
- Rubber tip
- *Advantages:* Lightweight, easily adjustable, freedom for hand activities
- *Disadvantages:* Need more strength, requires more skill and better trunk balance

Platform Crutches

- *Advantages:* Avoids weight bearing through wrist and hand (i.e., fractures, arthritis of wrist or hand, or weakness of triceps or grasp)
- *Disadvantages:* Awkward, heavy

🕮 WALKERS

Indications

1. Bilateral weakness and/or incoordination of the lower limbs or whole body
2. Whenever a firm, free standing aid is appropriate to increase balance (i.e., multiple sclerosis [MS] or parkinsonism)

3. To relieve weight bearing either fully or partially on an LE (allow the upper extremities to transfer body weight to the floor)
4. Unilateral weakness or amputation of the lower limb where general weakness makes the greater support offered by the frame necessary (i.e., osteoarthritis or fractured femur)
5. General support to aid mobility and confidence (i.e., after prolonged bedrest and sickness in the elderly)
   - *Advantages:*
     - Provide a wider base of support
     - More stable base of support
     - Provide a sense of security for patients fearful of ambulation
   - *Disadvantages:*
     - More conspicuous in appearance
     - Interferes with development of a smooth reciprocal gait pattern (e.g., ↓ step length with step-to-gait pattern)
     - Interferes with stair negotiation/difficult to maneuver through doorways or bathrooms
   - Types:
     - Lightweight walker
     - Folding walker
     - Rolling walker (two-wheel and four-wheel "rollator")
       - Two-wheel walker: More maneuverable and functional than a walker with no wheels
       - Rollator: Least stable and used for individuals who do not need frequent rest breaks or to fully offload lower limb (Sehgal, 2021)
     - Forearm resting walker
     - Hemi walker
   - 🕮 Base of support by assistive device: Single-point cane/single crutch < quad cane < hemi walker < two single-point canes/two crutches < rolling walker (Bradley & Hernandez, 2011)

SHOES AND LOWER LIMB ORTHOSES

SHOE COMPONENTS (FIGURE 6–13)

- A shoe consists of the upper, the sole, and in most cases an added heel.

Upper

- Part of shoe above the sole
- The shoe upper is made of a flexible material: Leather, woven fabrics, or synthetic materials, such as urethane or vinyl.
- In the **oxford style shoe** (also see next section), the *upper* consists of the following basic parts:
  - **Vamp** (one piece): Anterior part covering the instep and toes
  - **Quarters** (two pieces sewn together): The pieces that make up the posterior part; laterally, the quarter is cut lower to avoid infringing on the lateral malleolus
  - **High quarter:** Referred to as high top, offers ML stability
- **Tongue:** Strip of leather lying under the laces
- **Throat:** Opening at the base of the tongue, entrance to shoe
  - The more anteriorly the throat is located, the more room for internal modification (extra depth shoes also allow more room for shoe orthotics).
- **Toe box:** Anterior part of the vamp, protects toes from trauma

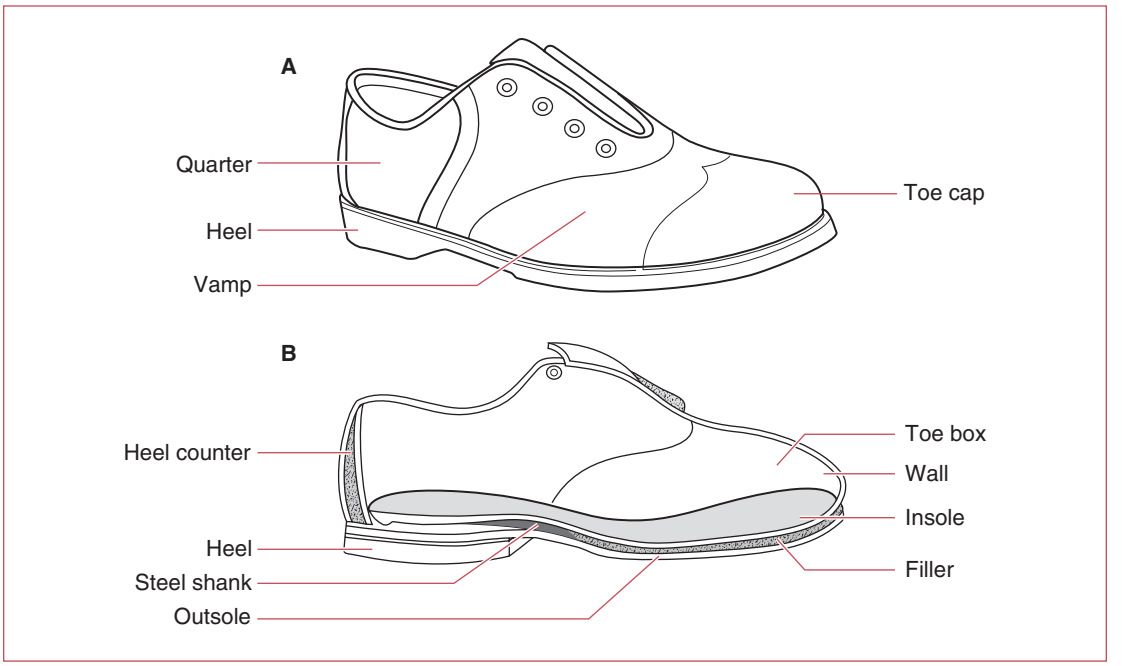


FIGURE 6–13 Oxford shoe component parts. (A) External view. (B) Longitudinal section.

Sole

- Bottom part of the shoe consisting of three layers: Insole, outsole, and the filler between them.
- **Outsole:** Part that touches the ground; may be made of leather, rubber, crepe, plastic, wood, or other materials
- **Insole:** Made of thin leather or man-made material; part closest to foot
- The **filler** is usually made of cork dust and latex.
- **Leather soles:** Indicated if shoe modifications are needed or attachment to metal AFO
- **Rubber soles:** Modifications are more difficult.
- **Ball:** Widest part of sole, at the metatarsal heads
  - Can be modified internally or externally to help alleviate forefoot pain
- **Shank:** Narrowest part of sole; between heel and ball
  - Usually reinforced with metal, leather, fiberboard, or other firm material (to provide additional support for the shoe in the region corresponding to the arch of the foot)
  - Can be reinforced with metal for attachment of metal AFO
- **External heel seat:** The posterior part of the sole to which the heel is attached

Heel

- Attached to outer sole
- The heel height varies from a negative heel that is lower than the forefoot position to 2 to 3 in. high
- Purpose of heel:
  - Acts as a shock absorber and prevents shoe from wearing out
  - Shifts weight to the forefoot
- Materials: Leather, wood, plastic, rubber, or metal
- Heel block: Part attached to heel seat, made of firm material
- Breast: Anterior part of heel; height of heel is measured in 1/8 in. at the breast
- **Types of heels:**
  - Flat heel: Broad base, measures 0.75 to 1.25 in. in height

  - Thomas heel: Flat with medial extension to support weak foot arch
  - Military heel: Slightly narrower base, measures 1.25 to 1.375 in.
  - Cuban heel: Still narrower base, higher heel
  - Spring heel: Placed under outer sole, eliminates breast, only 0.125 in.
- Many athletic shoes eliminate the heel because one can run faster this way.
- Height may be a factor in clinical conditions:
  - Shortening of gastrocnemius
  - Low back pain
  - High heels make ankle and foot more unstable (remember, the articular surface of talus is narrower posteriorly; this is the area of contact when the ankle is plantar flexed).
- **Heel counter:**
  - Reinforces the shoe, stabilizing the foot by supporting and controlling the calcaneus
  - Usually extends anteriorly to the heel breast
  - May extend further in specially made shoes
- **Collar:**
  - Band of leather stitched to the top of the quarters
  - Sometimes used to reduce positioning
  - Can also prevent shoe from falling off
- **Lace stay:**
  - Portion containing the eyelets for laces
  - Usually part of the vamp
  - Can be part of the quarters

BASIC OXFORD (LOW-QUARTER) SHOE TYPES

- **Blucher-style opening:** Front-laced shoe in which the quarters (specifically the lace stays) are not attached (stitched) distally to the vamp and remain loose and fully open, thus leaving more room for the entering foot
- **Bal or Balmoral-style opening:** Front-laced shoe in which the lace stays meet in front and are stitched to the vamp

SHOE MODIFICATIONS

Internal Modifications

- **Heel cushion-relief/excavation:** A soft pad with excavation under the painful part of the heel (e.g., a calcaneal spur)
- **Inner sole excavation/relief:** A soft pad with excavation under one or more painful bony prominences (usually metatarsal heads). The excavation is usually filled with compressible material.
- **Scaphoid pads/arch cookies/navicular pad:** A rubber, cork, or plastic foam wedge used to provide medial longitudinal arch support
  - Arch support insole: Used to support a weak long arch, increases stability (Del Toro & Seslija, 2019; Osborne & Esser, 2019)
- **Metatarsal pads:** Dome-shaped pads glued to the inner sole with the apex under the metatarsal shafts. It relieves pressure from the metatarsal heads by transferring the load to the metatarsal shafts.
- **Internal heel wedges:** Can be applied medially and promote hindfoot inversion (e.g., in flexible pes planus) or laterally, and promote hindfoot eversion and relive pressure on the cuboid (e.g., in flexible pes varus). It also increases total plantar-bearing area and can be used in fixed pes varus.
- **Toe crest:** Used to relieve pressure at the distal end and plantar surface of the toes (typically for hammer toes)

- **Foot orthoses (inserts or inlays):**
  - 🕮 **University of California Biomechanics Lab (UCBL) orthosis:** Custom-molded orthosis used to control flexible calcaneal deformities (rearfoot valgus or varus) and transverse plane deformities of the midtarsal joints (forefoot abduction or adduction)
    - It encompasses the heel and hindfoot and holds the midfoot with high medial and lateral trim lines.
    - It provides effective longitudinal arch support and realigns a flexible flat foot. It allows for subtalar supination, holds the calcaneus in place, and prevents subtalar pronation.
  - **Heel cup:** Provides calcaneal support; used to prevent lateral calcaneal shift (calcaneal valgus) in the flexible flat foot
  - **Longitudinal arch insert:** Can be applied medially or laterally
  - **Plastazote full-contact insoles:** Custom-molded, made of various densities and thickness. Accommodates many foot problems/deformities. Provides almost immediate comfort for plantar pain.
  - Other available foot orthoses include sesamoid inserts, metatarsal inserts, Levy balancer, Mayer, Morton's toe extension.

External Modifications

SOLE MODIFICATIONS

- **Rocker bar:** This is a convex strip placed across the sole just posterior to the metatarsal heads. It is longer than the metatarsal bar.
  - It can be used to relieve metatarsal pain (by relieving pressure), quicken the gait cycle (by assisting rollover during stance), or assist dorsiflexion or decrease demand on weak plantar flexors (push-off).
  - Rigid rocker-bottom shoes have also been recommended for use in cases of plantar fasciitis, neuropathic ulceration, and trauma to the midfoot or metatarsal region.
  - It can include the entire heel and sole to become a rocker-bottom shoe.
- **Metatarsal bar:** This is a simple bar placed in the sole just posterior to the metatarsal heads. It relieves pressure from the metatarsal heads by transferring load to the metatarsal shafts during stance.
- **Sole wedge:** A lateral sole wedge promotes forefoot eversion. A medial wedge promotes forefoot inversion.
- **Toe wedge:** A medial toe wedge is used to encourage toeing-in and is commonly used with a medial heel wedge. A lateral toe (pigeon-toe) wedge is used to assist the foot to toe out.
- **Sole flare:** Provides greater stability by widening the base of support of the shoe; a medial flare resists eversion, while a lateral flare resists inversion of the foot
- **Shank filler:** Applied medially to support medial longitudinal arch or laterally to support lateral longitudinal arch
- **Steel shank:** Inserted between the inner and outer soles to prevent motion of the anterior sole (bending of the sole) and thus reduce stress on metatarsals/phalanges. It is commonly used with the rocker bar to assist rollover.
- **Shoe lift:** Shoe modification that raises entire foot with rigid, noncompressible material. It is used for treating a leg length discrepancy of up to 2 cm. Shoe lifts ≥5cm are associated with increasing instability (Vogt et al., 2020).

HEEL MODIFICATIONS

- **Heel wedge:** Can be placed either medially (to rotate hindfoot into inversion) or laterally (to rotate hindfoot into eversion)
- **Heel flare:** Applied either medially (to resist hindfoot eversion) or laterally (to resist hindfoot inversion)

- **Heel extension:** Can project anteriorly either medially (Thomas heel) or laterally (reverse Thomas heel). The Thomas heel provides support to the medial longitudinal arch; the reverse Thomas heel provides support to the lateral longitudinal arch.
- **Cushioned heel ("SACH" heel):** A heel with the posterior portion replaced by a rubberized, compressible material to absorb shock at heel (foot) strike. Cushioned heel with shift weight line (line of gravity) anterior to the knee joint at initial contact, causing an extension moment of the knee (thus stabilizing knee)
- **Heel lift:** Used to compensate for fixed pes equinus deformity or for a leg length discrepancy of more than 1/4 to 1/2 in. (or if symptomatic)

SHOE MODIFICATION PRESCRIPTION AND FOOT ORTHOTICS

Insensate and Dysvascular Foot

CAUSES

- Peripheral neuropathies
- Diabetes
- PVD
- Peripheral nerve or spinal cord injury
- Myelodysplasias
- Stroke
- Chronic venous insufficiency

CAREFUL EVALUATION

- Assessment of skin condition.
- Evaluate distribution of plantar pressure during standing and walking.
- Evaluate thickness of fat pads, calluses, and scarring on the plantar surfaces.
- Sensory-impaired feet with no soft tissue or skeletal deformity need carefully fitted shoes and custom-molded foot orthotics.
- Thermocork and Plastazote insole will distribute stress over larger area.
- Extra depth shoe will be needed to accommodate the foot orthotics.

BONE DEFORMITY ON PLANTAR ASPECT

- Soft-molded insole of polyethylene foam recommended
- Areas of relief under bony prominences
- Metatarsal bar attached to flexible sole will relieve stress at metatarsal heads.
- When sole is rigid, rocker bar may be fitted.

PREVENTIVE FOOT CARE

- Regardless of cause, patient must follow daily foot care and inspection program.
- Patient may not be aware that footwear is too tight or breakdown is occurring.
- Daily foot examination
- Detect areas of tissue trauma
- Particular attention to metatarsal heads
- May need mirror
- Handicapped patient may need assistance.
- Skin may lack normal perspiration moisture.
- Skin may have scaling and fissuring.
- Daily foot soaks followed by application of emollient may be needed.

🕮 OFFLOADING SHOES

- Forefoot offloading shoes: Rigid sole design utilizing rocker or wedge design, which offloads the hallux and first metatarsal head to treat neuropathic ulcers and foot trauma (Fuchs et al., 2020)

- Hindfoot offloading shoes: Rigid rocker sole design to assist with healing plantar ulcers and postoperative healing (i.e., calcaneal fractures or ligament reconstruction; Mazur et al., 2019)
  - Increases midfoot load; avoid in individuals with midfoot injuries or diabetic disease
- Use of a unilateral offloading shoe elevates one limb, leading to decreased gait speed, decreased stride length, increased gait cycle time, increased gait unsteadiness, and limp (Ling et al., 2020).
- Compared with removable offloading shoe, a nonremovable total contact cast is more effective in ulcer healing (de Oliveira & Moore, 2015).

Arthritic Foot

GENERAL

- The foot is involved in 90% of patients with RA.
- Metatarsophalangeal (MTP) joints often affected early
- Tenosynovitis, rheumatoid nodules, and inflamed bursae are common.
- Atrophy of fat pads occurs.
- Advanced: Hallux valgus, clawing of toes, spread of forefoot, and rigid flat foot

SHOE PRESCRIPTION AND MODIFICATIONS

- Extra depth orthopedic shoes to accommodate custom-molded foot orthotic
- High toe box for hammer toes; wide toe box for bunion/hallux valgus
- Soft heel counter: Useful in patients with heel pain due to nodules or bursae
- Soft toe box: To adjust for deformities
- Metatarsal pad: Relief of pressure on painful metatarsal heads
- Full-length-molded insole: May be preferable to metatarsal pad that can move
- Redistribute weight over entire weight-bearing surface
- External modification with metatarsal bar may reduce pressure over MTP region.

CUSTOM-MADE SHOE

- Severely deformed foot will require a custom-made shoe.
  - Made according to plaster cast of the foot
- Leather should be used for the upper; sole can be leather or rubber.
- Other options include sandals with soft-molded insole and straps adjusted to avoid pressure areas.
- High heels are heavy and not suitable.
- May also require modification of closure as hands may be involved

Painful Foot

HEEL SPURS AND PLANTAR FASCIITIS

- Full-length-molded foot orthotics with insert of soft rubber or soft-grade polyethylene foam (Plastazote®) under heel
- Decrease in amount of weight bearing
- Support the arch to relieve stress on the plantar fascia

🕮 HALLUX RIGIDUS

- Degenerative joint disease (DJD) of the first metatarsal head
- Treatment goal is to reduce painful dorsiflexion and reduce pressure off joint.
- Treat with rocker-bottom sole, navicular pads, carbon fiber plate, and high toe box (Lam et al., 2017).

FOOT PROBLEMS ASSOCIATED WITH RUNNING

FACTORS

- Training errors: Most common cause (too much, too far, too fast)
- Poor flexibility

- Inadequate warming-up
- Biomechanical abnormalities
- Poor or worn footwear
- Growth and development
- Poor training surface

SPORTS SHOES

- All provide cushioning, stability, and surface traction.
- Varied designs to meet specific demands of individual or sport
- Remember—check the shoes!

PRONATED FOOT

- Most common biomechanical problem with runners
- Associated problems:
  - Tibial stress syndrome
  - Patellofemoral syndrome
  - Posterior tibial tendinitis
  - Achilles tendinitis
  - Plantar fasciitis

SHOE MODIFICATIONS

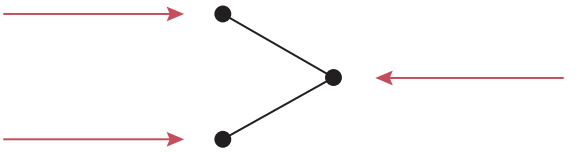
- Board and straight-last construction
- Motion control heel counter
- Medial support
- High-density insole medially
- Wider flared heel

SEMIRIGID FOOT ORTHOSIS

- Custom-molded thermoplastic foot orthotic with soft synthetic cover
- Medial wedge at the heel may be needed.
- For permanent use—made according to plaster cast of the foot

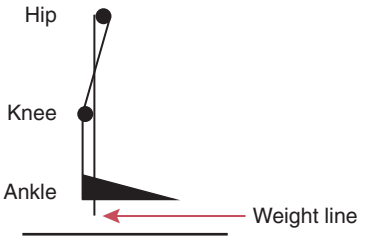
ORTHOTICS

- An orthosis, or brace, is an external device applied to body parts to provide one or more different functions, including:
  1. Reduction in pain/comfort
  2. Prevention or correction of deformity
  3. Support/stability
  4. Improvement in function
  5. Augmentation of weak muscles (assist motion)
  6. Control of spastic muscles
  7. Limitation of ROM (restriction of motion)
  8. Unloading of diseased or damaged joints
  9. A kinesthetic reminder (orthosis provides sensory/visual feedback that reminds the patient to adopt a more corrective or appropriate position, or to avoid some activities/movements)
- Biomechanical application of force and counterforce ⇒ three-point principle (see the following diagram box).



In all orthotic devices, three points of pressure are needed for proper control of a joint.

- 🕮 Line of gravity (weight line): Line passing through the COG to the ground (see the following diagram box):



🕮 During "quiet" standing, line of gravity (weight line) passes:
- Posterior to hip joint
- Anterior to knee joint
- Anterior to ankle joint

  - Line of gravity passes behind the cervical vertebrae, in front of the thoracic vertebrae, and behind the lumbar vertebrae.
  - Line of gravity is slightly posterior to the hip joint and tends to passively extend the hip joint.
  - Line of gravity is anterior to the knee joint and tends to passively extend the knee.
  - Line of gravity passes 1 to 2 in. anterior to the ankle joint and tends to dorsiflex the ankle. This motion is resisted by the soleus and gastrocnemius muscles.
- 🕮 While standing, the COG is in the midline and just anterior to the S2 vertebra.
- When selecting appropriate materials for orthotic devices, their strength, durability, flexibility, and weight need to be considered carefully.
- The orthotic design should be simple, inconspicuous, comfortable, and as cosmetic as possible.

MATERIALS USED IN ORTHOTICS

- **Metals**
  1. Steel—*Advantages*: Low cost, abundant, fatigue-resistant, provides high strength and rigidity. *Disadvantages*: Weight, need for expensive alloys to prevent corrosion
  2. Aluminum—*Advantages*: Corrosion-resistant, provides high strength, lightweight. *Disadvantages*: Lower endurance limit under repeated dynamic loading conditions than steel
  3. Titanium alloys—*Advantages*: Strength comparable to steel with only 60% of the density, more resistant to corrosion than aluminum or steel. *Disadvantages:* Limited availability, high cost
  4. Magnesium alloys—*Advantages*: Very lightweight, useful when bulk instead of strength is important
- **Leather**: Most commonly used as covering for braces and straps, covering for pelvic bands, and various types of molded applications, such as the girdle for the Milwaukee braces
- **Rubber**: Tough resiliency, shock-absorbing qualities. It can be used in padding for various assistive devices, seals in hydraulic mechanisms, and padding in body jackets and limb orthoses.
- **Plastics**
- 🕮 **Thermoplastics:** Soften and moldable when heated. It hardens when cooled, so it can be molded and remolded by heating.

1. Low-temperature thermoplastics:
   - Can be molded at temperatures just above body temperature (<80°C or <180°F); therefore, they can be shaped directly to the body without the need for a cast
   - Cannot be used effectively when high stress is anticipated, as in spasticity or in many LE applications. Main use is in upper limb orthotics.
2. High-temperature thermoplastics:
   - Used to manufacture permanent orthotic devices using vacuum-forming techniques
   - Major types: Acrylic, polyethylene, polypropylene, polycarbonate, ABS (acrylonitrile-butadiene-styrene), vinyl polymers, and copolymers
3. Thermosetting plastics: Develop a permanent shape when heat and pressure are applied and maintain a memory. More difficult to use than thermoplastics and generally cause more body irritation and allergic reactions. Examples: Polyesters, epoxies, and polyurethane foam.
4. Carbon fiber—*Advantages*: Lightweight, high strength. *Disadvantages*: Very expensive, difficult to shape or modify

LOWER LIMB ORTHOTIC PRESCRIPTIONS (ANKLE–FOOT ORTHOSIS, KNEE–ANKLE–FOOT ORTHOSIS, HIP–KNEE–ANKLE–FOOT ORTHOSIS)

Ankle–Foot Orthoses

An **ankle–foot orthosis (AFO)** is commonly prescribed for weakness affecting the ankle and subtalar joints. These can be made of plastic, metal, carbon, or a hybrid.

PLASTIC ANKLE–FOOT ORTHOSES

- Plastic AFOs are either prefabricated out of thermoplastic or custom-molded over a model of the patient's limb taken from a casting. Generally, they would encompass the posterior calf area and continue down posteriorly by the ankle and extend down the plantar surface of the foot to assist in dorsiflexion and limit plantar flexion, with anterior Velcro strap closure at proximal calf.
- The design of the **trim lines** and the design of the foot plate will help determine the structural support and rigidity of the device. The following includes typical trim line designs:
  1. **PLS:** Most flexible plastic AFO with a very thin plastic band behind the ankle that allows the patient to overpower the brace during the push-off/plantar flexion phase but also provides dorsiflexion assist and positioning of the foot during the swing phase. This design is typically used for *flaccid foot drop*.
  2. **Semirigid plastic AFO:** Trim line is just behind the malleoli and provides increased ML stability of the ankle and dorsiflexion positioning of the foot. Less motion is allowed with this brace design, and the patient cannot easily propel during push-off. This design is most commonly used for patients who have a foot drop with some extensor tone and/or ML instability of the ankle.
  3. **Rigid (solid) plastic AFO:** Trim line at the malleoli or anterior to the malleoli with no motion allowed at the tibiotalar or subtalar joint. Commonly used for patients with the highest levels of spasticity/tone or when complete immobilization of the ankle is necessary. The standard solid plastic AFO would typically have a foot plate extending through the metatarsal heads (three-fourth-length foot plate), but if the toes are spastic and claw into a flex position a full foot plate should be used. Inhibitory foot plate designs are used, which put the toes into extension to help reduce tone throughout the entire limb. This brace design is typically used for patients with the highest levels of spasticity, early to moderate Charcot joint, and for postoperative immobilization of the foot or ankle.

PLASTIC VERSUS METAL ANKLE–FOOT ORTHOSES

Plastic design AFOs are most desirable due to their lightweight, intimate fit, cosmetic appeal, and lack of attachment to the shoe. However, in certain instances, the selection of plastic materials may be inappropriate, and a metal AFO should be considered:

- Risk of excessive pressure on or presence of skin breakdown on the leg or foot
- Patients with insensate foot due to peripheral neuropathy or peripheral nerve injury should be considered candidates for metal AFOs rather than plastic.
- Patients with fluctuating edema that is not managed with compression stockings should be considered most appropriate for metal AFOs rather than plastic AFOs.

ANKLE–FOOT ORTHOSES WITH HINGED (ARTICULATED) ANKLE JOINTS

- In addition to preventing ML instability, most mechanical metal ankle joints either control or assist dorsiflexion and plantar flexion by means of stops (pins) or assists (springs).
- *Indications/advantages:*
  1. Paralysis of dorsiflexion, plantar flexion, foot inversion, and foot eversion
  2. Prevention and correction of deformities
  3. Reduce the energy cost of ambulation—for example, patients with spastic diplegia (e.g., cerebral palsy), lower motor neuron weakness (e.g., poliomyelitis), and spastic hemiplegia (e.g., stroke)
- 🕮 **Single-channel ankle joints** have three options:
  - Inserting a spring in the channel for dorsiflexion assist (Klenzak joint)
  - Inserting a steel pin for plantar flexion stop
  - Inserting both a pin and a spring for dorsiflexion assist and plantar flexion stop
- 🕮 **Dual-channel ankle joints** have both posterior and anterior channels. The posterior channel functions exactly the same as a single-channel joint described earlier. The anterior channel provides the additional option of an adjustable steel pin to block the forward progression of the tibia at midstance (dorsiflexion stop), or to lock the joint in a fixed position. This can be useful when the quadriceps muscle is weak or when there is Charcot joint deformity at the ankle.

Knee–Ankle–Foot Orthoses

1. A knee–ankle–foot orthosis (KAFO) represents an extension of the AFO proximally to control knee motion and alignment. It comprises an AFO with bilateral uprights, a knee joint, and two thigh bands. A KAFO may have a *single axis* (most common) or *polycentric* type of knee joint.
2. The following are different types of **single-axis knee joints**:
   - A *free motion knee joint* provides unlimited flexion but usually has a stop to prevent hyperextension. Used for patients with genu recurvatum but with enough strength to control the knee in stance and ambulation.
   - An *offset knee joint* places the hinge posterior to the knee joint so the patient's weight line falls anterior to the offset joint, stabilizing the knee during the early stance phase. It is free to flex during the swing phase and allows sitting without the need to manipulate the lock.
     - It should not be used in patients with knee or hip flexion contractures, or with a plantar flexion stop at the ankle. The patient must be careful when walking on a ramp as the knee may flex inadvertently.
     - 🕮 KAFO with double metal uprights and a posterior offset knee joint would be a good option for an obese patient with quadriceps weakness.
   - A *trick knee* allows 0- to 25-degree movement while in the locked position to normalize gait. Free knee when unlocked to allow sitting.
   - *Ratchet knee joints* are usually used to stretch out knee flexion contractures. The rachet joint locks every 7 to 10 degrees, from 90 degrees of flexion to full extension.

- An *adjustable knee lock joint (dial lock)* has a serrated adjustable knee joint that permits locking in almost any degree of flexion. It is useful in patients with a knee flexion contracture that is diminishing with treatment.
- *Trigger lock knee joints* prevent knee buckling. To be able to lock the joint, the patient must be able to fully extend the knee, either actively or passively. Contraindicated in patients with knee contracture.
- *Drop lock knee joints* have rings that drop over the joints in extension, locking them. They are designed to drop either by gravity or with assistance from the patient.
- A *bail lock* engages when the knee is fully extended; there is a semicircular lever (bail) attached posteriorly that unlocks the joint with an upward pull on the bail (either manually or by backing up to sit down on a chair).
  - Disadvantages: The bail is bulky and can be accidentally released if it hits a rigid object.

3. A **polycentric knee joint** provides unlimited knee flexion and extension but has significant added weight and components that require maintenance.

🕮 SCOTT-CRAIG ORTHOSIS (SCOTT-CRAIG LONG LEG BRACE; FIGURE 6–14)

- Bilateral KAFOs are designed for standing and ambulation in adults with paraplegia. They provide patients with a complete neurologic level at L1 or lower with a more functional and comfortable gait.
- It eliminates unnecessary hardware to reduce weight and facilitate donning/doffing (eliminates the lower thigh and calf band).
- Components:
  1. Sole plate extending to the metatarsal heads with a crossbar added to the metatarsal heads area for ML stabilization
  2. Ankle joint set at 10 degrees of dorsiflexion
  3. Anterior rigid tibial band (patellar tendon strap)
  4. Offset knee joint with bail lock
  5. Proximal posterior thigh band
- 🕮 *Unsupported standing* is possible with Scott-Craig orthoses.
- With the ankles and knees locked, hip stabilization can be achieved by leaning the trunk backward so that the COG of the trunk rests posterior to the hip joint, resulting in tightening of the anterior hip capsule and the "Y" ligament. In adults, the "Y" ligament (iliofemoral ligament, ligament of Bigelow) is usually strong/stable enough to provide hip-pelvic stability when using a KAFO without pelvic bands.
- Paraplegic patients can ambulate with Scott-Craig orthoses and crutches or walker using a swing-to or swing-through gait pattern.

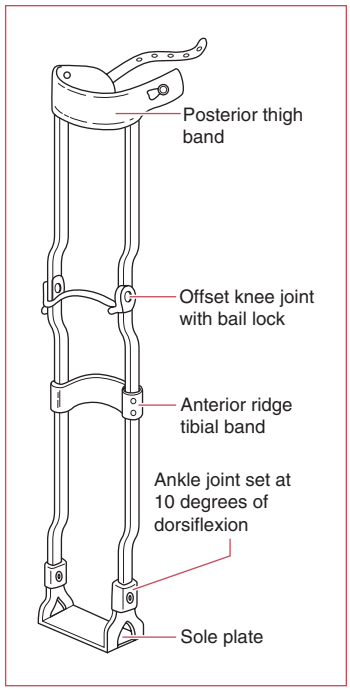


FIGURE 6–14 Scott-Craig orthosis.

Hip–Knee–Ankle–Foot Orthosis

- A hip joint and pelvic band attached to the lateral upright of a KAFO convert it to a hip–knee–ankle–foot orthosis (HKAFO).
- *Indications*:
  - Hip flexion/extension instability
  - Hip adduction/abduction weakness
  - Hip internal rotation/external rotation instability
  - Complete paralysis of the leg

🕮 RECIPROCAL GAIT ORTHOSIS

- One special design of HKAFO is the *reciprocal gait orthosis* (RGO). The RGO is used for upper lumbar paralysis in which active hip flexion is preserved.
- An RGO consists of bilateral HKAFOs with offset knee joints with drop locks, posterior plastic AFOs, thigh pieces, custom-molded pelvic girdle, hip joints, and a thoracic extension with Velcro straps in addition to the control mechanism.
- Several designs are available, including cord and pulley design (early versions), gear-box cable, single cable, dual-cable, and *isocentric RGO* (IRGO; latest design). In IRGO, the cord is substituted by a pelvic band attached to the posterior surface of the molded thoracic section (**Figure 6–15**).
  - *Advantages of the IRGO*: Less bulky appearance (no protruding cables in the back), may be more cost-efficient than cable RGO (no energy loss due to cable friction)
- In all RGOs, the hip joints are coupled together with cables (or to the pivoting pelvic band in the IRGO), which provides mechanical assistance to hip extension while preventing simultaneous bilateral hip flexion.
- As a step is initiated and hip flexion takes place on one side, the cable coupling induces hip extension on the opposite side, producing a reciprocal walking pattern.
- Forward stepping is achieved by active hip flexion, lower abdominal muscles, and/or trunk extension.
- Using two crutches and an RGO, paraplegics can ambulate with a four-point gait. A walker may also be used.

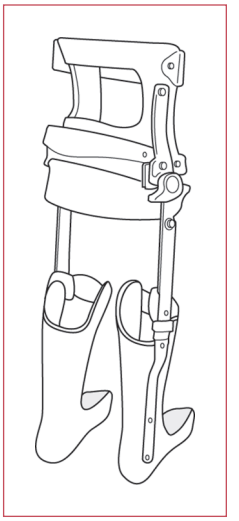


FIGURE 6–15 Isocentric type RGO.
RGO, reciprocal gait orthosis.

Knee Orthoses

- A knee orthosis provides support to the knee joint. The type of support it gives depends on the plane of motion that is being limited.
  - Sagittal plane: Typically limits hyperextension of the knee (genu recurvatum)
  - Axial rotation: Provides ML and axial control
  - Coronal plane: Theoretically mimics anatomic knee joint function
- Prescribed to prevent genu recurvatum and to provide ML stability. Prophylactic knee orthoses, theoretically, are designed to prevent injury to the knee or at least to reduce the degree to which the knee is injured in athletic activities.
- Knee orthoses may be used during sports and other physical activities to provide functional support for unstable knees or during rehabilitation after injury or surgery of the knee. The use of knee orthosis for the prevention of knee injuries in sports is controversial.
- Numerous designs of knee orthosis are available. Most consist of two uprights, free or adjustable knee joints, as well as thigh and calf cuffs.
- 🕮 Adding a foot plate to a knee immobilizer decreases the rotational instability of the knee.

FLEXIBLE KNEE ORTHOSES

- Made from an elastic material or rubber
- May include hinged metal knee joints, patellar pads, adjustable straps to change tension, and an anterior cutout to relieve pressure on the patella when the knee is flexed.
- Functions:
  - Provide comfort for patients with osteoarthritis, minor knee sprains, and mild edema
  - Proprioceptive feedback/kinesthetic reminder
  - Minimal mechanical support

- Can retain body heat
- Theoretically stabilizes patellar tracking with patellofemoral dysfunction
- 🕮 Hinged knee brace for grade II and III MCL injuries, worn for 3 to 6 weeks (Vosoughi et al., 2021)

KNEE ORTHOSES—FORCE SYSTEMS

TO LIMIT FLEXION:
1. Anteriorly directed force on posterior proximal thigh 2. Posteriorly directed force on anterior surface of the knee (either directly over the patella or a combination of support just proximal and just distal to the patella) 3. Anteriorly directed force on posterior aspect of the calf
TO LIMIT EXTENSION (GENU RECURVATUM):
1. Two bands placed anterior to the knee axis (one superior and one inferior to the knee) 2. One band placed posterior to the knee joint in the popliteal area 3. It also has an additional thigh band with longer uprights to obtain better leverage at the knee joint.

TYPES OF KNEE ORTHOSES

- *Swedish knee cage* (**Figure 6–16**)**:**
  - Prefabricated brace that controls minor to moderate knee hyperextension/genu recurvatum due to ligamentous or capsular laxity
  - Available in nonarticulated and articulated forms
  - The articulated version prevents hyperextension but permits full flexion of the knee, using a three-point pressure system with two anterior straps and one posterior strap held in position by a metal frame medially and laterally (see earlier table).

FIGURE 6–16 Swedish knee cage.

FIGURE 6–17 Lenox-Hill orthosis.

- *Lenox-Hill derotation orthosis* (**Figure 6–17**)**:** Knee orthosis designed for control of knee axial rotation in addition to anterior-posterior and medial-lateral control
  - Used for protection and management of sports injuries to the knee (e.g., anterior cruciate ligament [ACL] injuries)
- 🕮 *Offloader knee brace:* Knee orthosis designed to realign the knee to redistribute knee compartment pressure, decrease pain, and improve stability (Ramsey & Russell 2009)
  - Valgus offloader brace realigns the knee in moderate to severe medial knee osteoarthritis (Ramsey & Russell 2009).

Knee Immobilizer

- Indications include acute or presurgical management of quadriceps or patellar tendon rupture, medial collateral ligament rupture, and patellar fracture or dislocation (Gravlee & Van Durme, 2007).

LOWER EXTREMITY ORTHOSES FOR PRESSURE REDISTRIBUTION

Patella Tendon Bearing Orthosis

- Reduces up to 50% of weight transmission through the mid or distal tibia, ankle, and foot
  - Examples: Healing of os calcis fracture, postoperative ankle fusion, heel with refractory pain, delayed union or nonunion of fractures or fusions, avascular necrosis (AVN) of the talar body, DJD of the talar or ankle joint, osteomyelitis of the os calcis, diabetic ulceration of the plantar surface, and Charcot joint
- Supports weight on the patellar tendon, calf muscle, or tibial flares with the load being transmitted to the shoe via metal uprights
- May have plastic bivalve design or calf corset if the patient has fluctuating edema
- Because little or no ankle motion is allowed, a cushion heel or a rocker bottom is added to provide smoother gait pattern.

Ischial Weight-Bearing Orthosis

- Quadrilateral brim or an ischial (Thomas) ring to relieve weight from the femur or knee

Patten-Bottom Orthosis

- Uses uprights with no ankle joint that terminate in a floor pad distal to the shoe so the foot is freely suspended in midair
- A shoe lift is needed on the opposite side to equalize leg length.

Fracture Orthosis

- Stabilizes the fracture site and helps promote callus formation by allowing weight bearing and joint movement after initial rest period to allow pain and edema to subside. Minimizes joint stiffness and reduces complications such as nonunions. Circumferential compression of the soft tissue can be used to prevent undue bony motion at the fracture site.

LOWER EXTREMITY TONE-REDUCING ORTHOSES (FIGURE 6–18)

Rationales for Efficacy of Tone-Reducing Orthoses

1. Inhibition of reflexes
2. Pressure over muscle insertions
3. Active and static prolonged stretch
4. Orthokinetics

1. INHIBITION OF REFLEXES

- A reflex consists of a motor act that is elicited by some specific sensory input.
- Primitive reflexes appear at birth and become integrated once more complicated movements emerge.
- When the central nervous system (CNS) is damaged, primitive reflexes reemerge and again dominate motor activity.
- Examples:
  - Toe (plantar) grasp reflex—*Trigger:* Pressure over ball of foot. *Response*: Increased tone in toe flexion and ankle plantar flexion. AFO design reportedly reduces stimulus pressure.
  - Inversion reflex—*Inversion trigger*: Pressure to medial border of foot over first metatarsal head
  - Eversion reflex—*Eversion trigger*: Pressure to lateral border of foot over fifth metatarsal head. AFO design reduces abnormal tone by stimulating antagonist reflex.

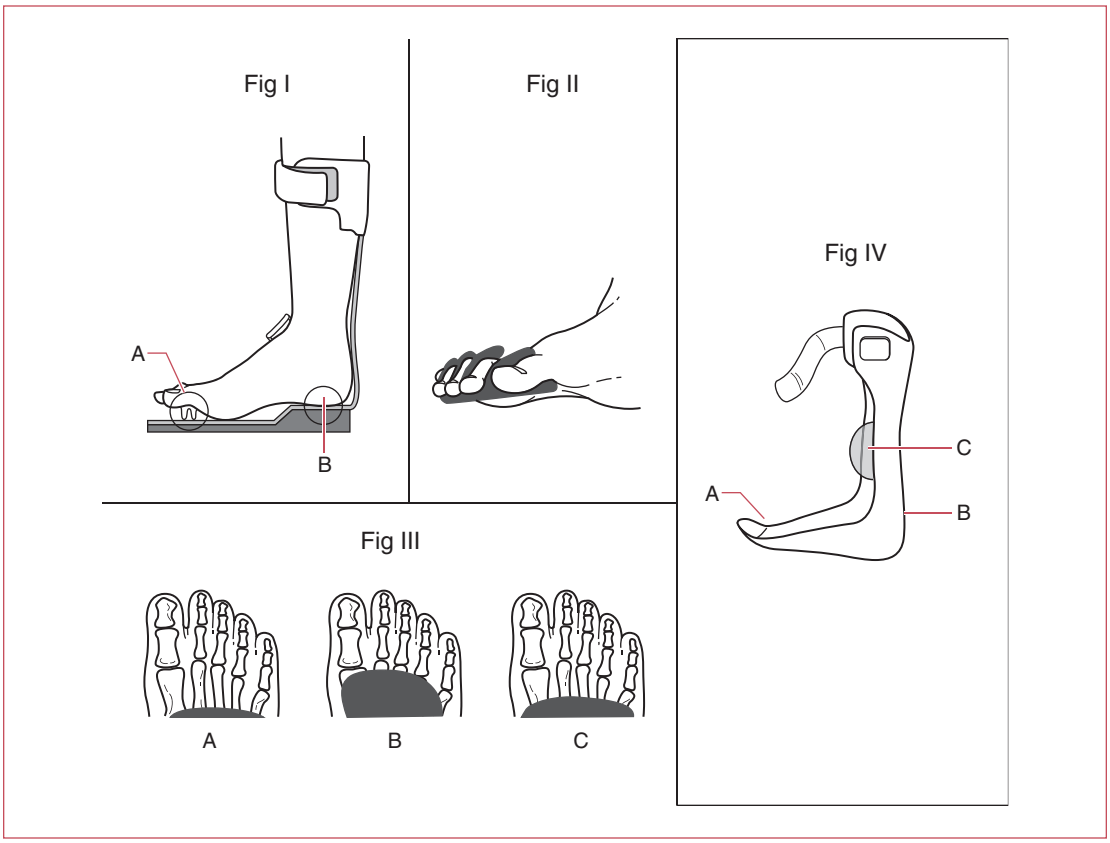


FIGURE 6–18 Tone-reducing orthoses: (I) Side view of tone inhibitory orthosis. (A) Spastic inhibitory bar. (B) Internal heel. (II) The neurophysiologic AFO is designed to provide total surface contact. The toe separators are made of Plastazote and facilitate toe extension. (III) Strategies using metatarsal pad placement. (A) Proximal to metatarsal heads. (B) Under metatarsal heads. (C) Medial extension to induce inversion reflex. (IV) Medial view of orthosis design to incorporate tone inhibitory characteristics of plaster casting. (A) Toe hyperextension plate. (B) Tendon pressure over gastroc-soleus insertion. (C) Lateral flange (for tone).
AFO, ankle–foot orthosis.

2. PRESSURE OVER MUSCLE INSERTION

- Farber reported in 1974 that continuous firm pressure at point of insertion reduces tone.
- AFO design: Pressure on either side of tendo calcaneus and insertion of gastroc-soleus muscle groups

3. ACTIVE AND STATIC PROLONGED STRETCH

- Decrease reflex tone by providing mechanical stabilization of the joint and altering properties of the muscle spindle
- AFO designs that provide total ankle–foot contact (**Figure 6–19**)

4. ORTHOKINETICS

- Originally developed in 1927 by Julius Fuchs, an orthopedic surgeon
- Focuses on physical effects to materials placed over muscle bellies
- Passive field materials (those that are cool, rigid, and smooth) produce an inhibitory effect.
- Active field materials (those that are warm, expansive, and textured) produce a facilitatory effect.
- AFO design:
  - Active field stimulation (e.g., foam) over anterior tibialis to encourage dorsiflexion
  - Passive field inhibition over gastrocnemius to reduce spastic plantar flexion
  - Dual orthokinetic concepts interrelated and applied simultaneously

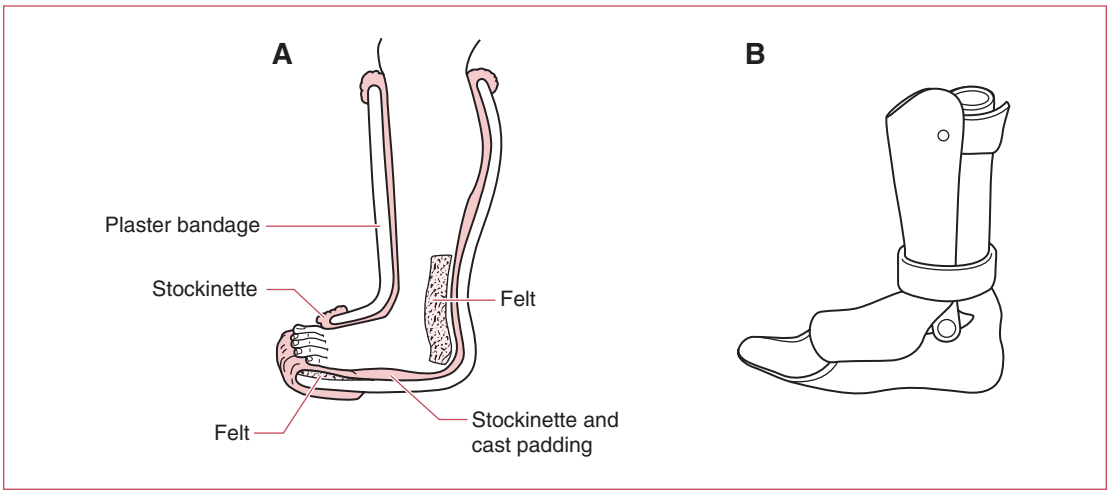


FIGURE 6–19 (A) Cross-section of inhibitory cast that provided heel and forefoot alignment on neutral as well as total surface contact. (B) Bivalved Chattanooga articulating orthosis provides uniform contact of inhibitory cast as well as adjustable ankle joint to maintain consistent static force.

UPPER LIMB ORTHOSES

- Upper limb orthoses may be categorized into the following types:
  1. Static orthoses
  2. Dynamic orthoses
  3. Tone-reducing orthoses

STATIC UPPER LIMB ORTHOSES

Indications for Static Orthosis Use

1. Immobilize, stabilize, and support a joint in a desired position
2. Protect weak muscles from overstretch
3. Prevent contractures
4. Support structures following surgical repair
5. Facilitate the healing of soft tissue injuries and fractures

Complications Associated With Static Orthosis Use

1. Skin breakdown
2. Contractures
3. Infection

Positional Orthoses

BASIC OPPONENS ORTHOSES

- 🕮 ***Opponens orthosis*** **(Figure 6–20)**: Primarily used to immobilize the thumb and first MCP joint to promote tissue healing and/or protection or for positioning of the weak thumb in opposition to other fingers to facilitate three-jaw chuck pinch

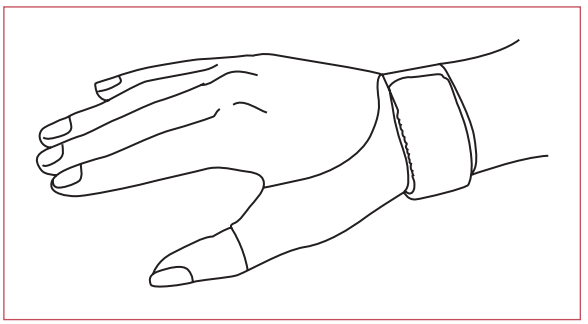


FIGURE 6–20 Basic opponens splint.

- Hand–thumb orthoses consist of a dorsal and a palmar bar encircling the hand, with a thumb abduction bar and C-bar to stabilize the thumb.

🕮 LONG OPPONENS ORTHOSIS WITH WRIST-CONTROL ATTACHMENTS

- Similar to short opponens orthosis but crosses the wrist. Stabilizes first MCP while forearm bar maintains wrist in extension and prevents radial and ulnar-deviation deformities.
- *Examples:* Long opponens splints (**Figure 6–21**) and thumb spica splints

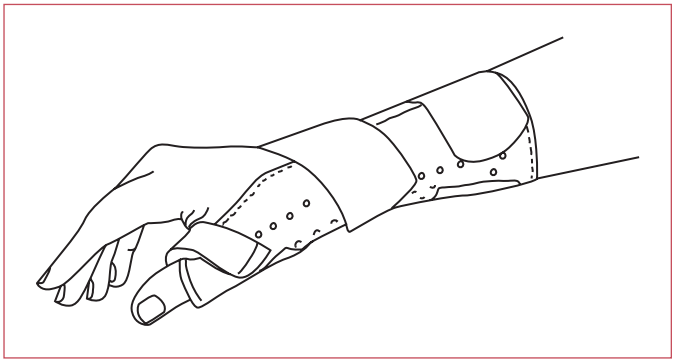


FIGURE 6–21 Long opponens splint (forearm, wrist, thumb).

OPPONENS ORTHOSIS WITH LUMBRICAL BAR

- Hand–finger orthosis (HFO) that prevents MCP joint hyperextension but allows full MCP flexion
- Prevents claw hand deformity (in addition to the benefits already mentioned for opponens orthoses)

OPPONENS ORTHOSIS WITH FINGER EXTENSION ASSIST ASSEMBLY

- Similar to basic opponens orthoses but adds outriggers that assist PIP and DIP extension
- Used for interphalangeal (IP) flexion contracture, boutonnière deformity, or postsurgical release of Dupuytren's contracture

UTENSIL HOLDER/UNIVERSAL CUFF (SPLINT)/ADL SPLINT (FIGURE 6–22)

- Consists of a handcuff with palmar pocket onto which a utensil can be inserted

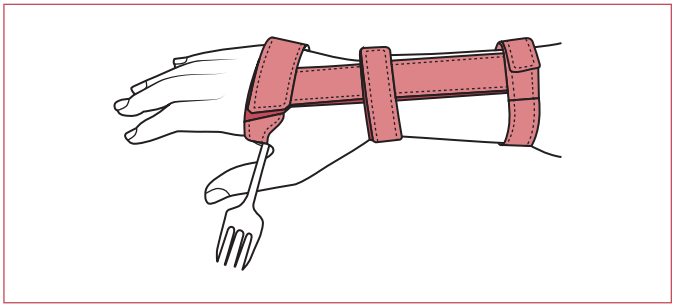


FIGURE 6–22 Utensil holder/universal cuff (splint).

FINGER STABILIZERS/STATIC FINGER ORTHOSES

- IP orthoses (DIP, PIP, and DIP + PIP gutter splints, static finger splints, aluminum and foam splints)
- Finger orthoses are used to restrict motions at the PIP and DIP.
- Generally, IP maintained in full extension to keep the collateral ligaments stretched and to prevent IP flexion contracture (unless condition dictates otherwise)
- Used to promote healing (e.g., phalanx fracture, PIP/DIP dislocation) and to provide prolonged finger stretch (e.g., burns and contractures)

RING ORTHOSES (FIGURE 6–23)

- Swan neck ring splint: Prevents PIP joint hyperextension through a three-point pressure system but allows full IP flexion
- Boutonnière ring splint: Immobilizes the PIP in extension and prevents flexion through a three-point pressure system

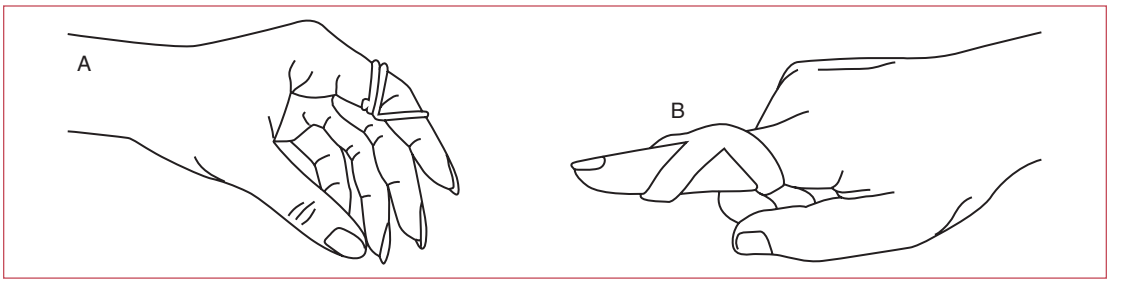


FIGURE 6–23 Ring stabilizers: (A) Swan neck splint. (B) Boutonnière splint.

Hand–Finger Orthoses

METACARPOPHALANGEAL ULNAR-DEVIATION RESTRICTION ORTHOSIS (FIGURE 6–24)

- Used to limit ulnar deviation of the MCP with unrestricted (if possible) MCP flex/extension in arthritic patients with ulnar deviation at the MCPs

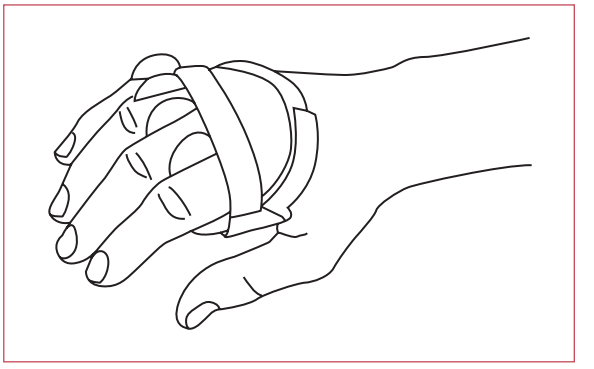


FIGURE 6–24 Ulnar-deviation correction splint.

Thumb Orthoses

THUMB CARPOMETACARPAL STABILIZERS/THUMB POSTS

- Thumb orthosis that stabilizes the first carpometacarpal (CMC) and MCP joints in neutral position to protect the thumb from inadvertent motion

THUMB-WEB SPACE STABILIZERS/THENAR WEB SPACERS/C-BAR SPLINT (FIGURE 6–25)

- HFO that consists of a rigid C-shaped splint held firmly in the thumb and index finger web space
- Function: Increases or maintains the thenar space and prevents web-space contractures
- Uses: Burns, postsurgical revision of scar, web-space contractures

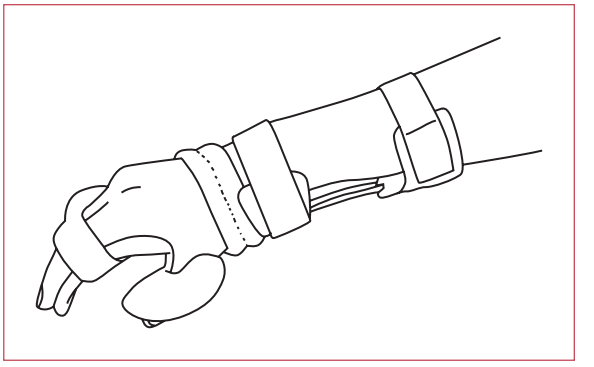


FIGURE 6–25 C-bar splint (thenar web spacer).

Wrist–Hand–Finger Orthoses

RESTING HAND SPLINTS

- Static wrist–hand–finger orthosis (WHFO) used to immobilize the wrist, fingers, and thumb
- Usually applied to the volar surface (can be dorsally applied or circumferential)
- Extend from the fingertips to two-thirds of the distal forearm
- Wrist in neutral to slight extension and the digits in an intrinsic plus position (MCPs flexed 70–90 degrees, IPs in full extension, thumb CMC in palmar abduction, and thumb MCP/IP in full extension)
- Immobilization in this position preferred because MCP joint and IP collateral ligaments are kept stretched, minimizing future joint capsule contractures
- Provide functional thumb position for opposition and three-jaw chuck pinch

Wrist–Hand Orthoses

VOLAR WRIST–HAND ORTHOSIS (WRIST COCK-UP SPLINT)

- Wrist–hand orthosis (WHO) extending from the distal two-thirds of the forearm to just proximal to MCP joint to allow full MCP flexion while maintaining the functional position of the wrist and hand
- *Uses*: Resting wrist and hand in acute arthritis (RA), wrist sprain/contusion, flexor/extensor tendinitis, carpal tunnel syndrome, postsurgical wrist extensor tendon repair, wrist fusion, skin grafting, contracture prevention, reduce pain, reduce spasticity, prevent ulnar/radial deviation of the wrist/hand (e.g., RA)

DORSAL WRIST–HAND ORTHOSES

- Provide the same functions of the volar WHO with greater stabilization because of rigid dorsal hand section
- More difficult to fabricate and fit than the volar WHO

DYNAMIC (FUNCTIONAL) ORTHOSES

Finger Orthoses

THUMB MOBILIZATION ORTHOSES

- *Thumb extension-mobilization orthosis*: Dynamic thumb IP extension splints. Used for thumb IP flexion contracture.
- *Thumb flexion-mobilization orthosis*: Dynamic IP flexion splints. Used for thumb IP extension contracture.
- *Thumb abduction-mobilization orthosis*: Dynamic thumb abduction splint. Used for thumb-adduction contracture.

Finger Mobilization Orthoses

- *IP extension-mobilization orthoses* passively extend the PIP joints.
  - *Uses*: Finger IP flexion contracture, boutonnière deformity, and postsurgical release of Dupuytren's contracture
  - *Examples*: Dynamic IP extension splints, reverse finger-knuckle benders, Capener splints, spring coil assist, eggshell finger extension casts, buddy splints
- *IP flexion-mobilization orthoses* passively flex PIP joints.
  - *Use*: Finger IP extension contracture
  - *Examples*: Dynamic IP flexion splints, finger-knuckle benders, fingernail hook orthoses, buddy splints

Metacarpophalangeal Mobilization Orthoses

- *MCP extension-mobilization orthoses*: Extend MCP joints
  - *Uses*: Extend MCP joints in MCP-flexion contractures, burns, and post-open reduction and internal fixation (ORIF) of metacarpal fracture, as well as patients with weak finger extension (e.g., radial nerve or brachial plexus lesion)
  - *Examples*: Reverse MCP knuckle benders, dynamic MCP extension splints with dorsal outrigger, MCP extension assists, radial nerve splints
- *MCP flexion-mobilization orthoses*: Flex MCP joints
  - *Uses*: Flex MCP joints in MCP collateral ligament contractures, extensor tendon shortening, median/ulnar lesion, claw hand, postcapsulotomy, post-ORIF of metacarpal fracture
  - *Examples*: MCP knuckle benders, dynamic MCP-flexion splints with volar outrigger and fingernail hooks, MCP-flexion assists

Wrist Orthoses

WRIST-CONTROL ORTHOSIS

- Promotes slight extension of the wrist or prevents wrist flexion, thus assisting weak grasp (via tenodesis effect)

VOLAR WRIST FLEXION-CONTROL ORTHOSIS (COCK-UP SPLINTS)

- WHOs in which the palmar section is extended (usually 0–20 degrees)
- Used to tighten finger flexors (via tenodesis effect) and prevent wrist flexion contracture in patients with radial neuropathy

WIRE WRIST-EXTENSION ASSIST ORTHOSIS (OPPENHEIMER SPLINT)

- Prefabricated from spring steel wire and padded steel bands to assist wrist extension by tensing the steel wire, thus aiding finger flexion through tenodesis effect

🕮 WRIST-DRIVEN PREHENSION ORTHOSIS (TENODESIS ORTHOSIS, FLEXOR HINGE SPLINT; FIGURE 6–26)

- Used in C6 complete tetraplegia (no muscles to flex or extend; fingers remain innervated, but wrist extension, through the extensor carpi radialis muscle, is intact) to provide prehension through tenodesis action and maintain flexibility of the hand, wrist, and elbow
- Wrist extensors should be 3+ or better to use body-powered tenodesis.
- The proximal and distal IP joints of fingers two and three and the CMC and MCP joints of the thumb are immobilized.
- May interfere with manual wheelchair propulsion
- Rarely accepted by C7 and C8 tetraplegics who prefer to use their residual motor power or utensil holders

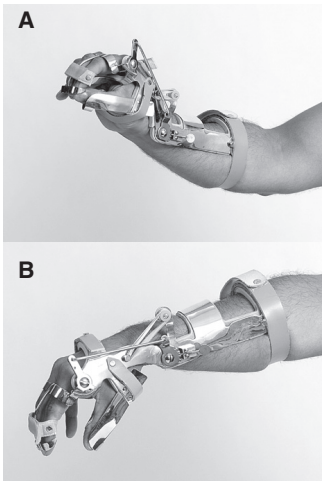


FIGURE 6–26 Wrist-driven prehension orthosis. (A) Wrist extension: Three-jaw chuck. (B) Wrist flexion: Release.

🕮 REHABILITATION INSTITUTE OF CHICAGO TENODESIS SPLINT (FIGURE 6–27)

- Made of low-temperature thermoplastics in three separate pieces (wristlet, short opponens, and dorsal plate over index and middle finger)
- Easily and quickly fabricated; made as a training and evaluation splint for patients; lightweight
- Uses a cord/string running from the wrist piece, across the palm and up between the index and ring fingers. The string is lax when the wrist is flexed and tightens with wrist extension, bringing the fingers close to the immobilized thumb, accomplishing three-jaw chuck prehension.

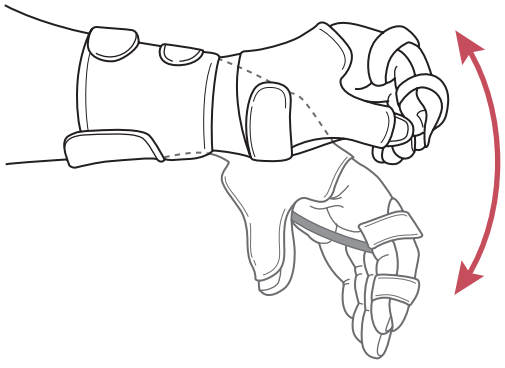


FIGURE 6–27 Rehabilitation Institute of Chicago (RIC) tenodesis splint.

Forearm Orthoses

🕮 BALANCED FOREARM ORTHOSIS (FIGURE 6–28)

- Shoulder–elbow–wrist–hand orthosis (SEWHO) that consists of a forearm trough (attached by a hinge joint to a ball-bearing swivel mechanism) and a mount (which can be mounted on the wheelchair, on a table or working surface, or onto the body jacket)
- Helps support the forearm and arm against gravity and allows patients with weak shoulder and elbow muscles to move the arm horizontally and flex the elbow to bring the hand to the mouth (e.g., patients with spinal cord injury, Guillain–Barré syndrome, polio, muscular dystrophy, and brachial plexus injury)

- *Requirements:*
  - Some residual muscle strength of biceps and pectoralis (manual muscle testing [MMT] at least poor or grade 2) and coordination of elbow flexion (can be used for C5 tetraplegic)
  - Adequate trunk stability and balance
  - Adequate endurance in a sitting position
  - Preserved ROM of the shoulder and elbow joints
  - Other uses: May be used in spastic patients to allow self-feeding by dampening muscle tone through a friction device

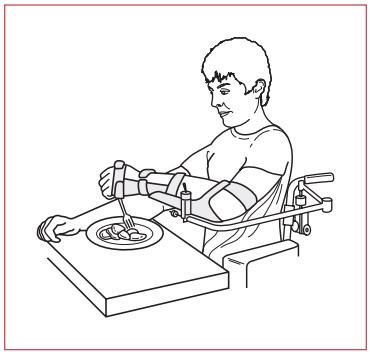


FIGURE 6–28 Balanced forearm orthosis.

Elbow Orthoses

- Assist in elbow flexion or extension
- Dynamic elbow splints, static progressive elbow splints, turnbuckle elbow splints—gently elongate the soft tissues over a long period in an attempt to reverse joint malalignment (contractures, burns, late phase of fracture). They are not used in spastic muscles, as they may further increase tone.
- A **dorsal elbow-extension-mobilization orthosis** extends the elbow and provides ML elbow stability and rotational forearm stability.
- A **dorsal elbow-flexion mobilization orthosis** flexes the elbow and provides ML stability and rotational forearm stability.

TONE-REDUCING ORTHOSES (FIGURE 6–29)

- Can be either hand-based orthoses (e.g., hand–cone splints) or forearm-based WHFOs (e.g., antispasticity ball splints, hand–cone–forearm splints).
- Can be volar-based, dorsal-based, or circumferential
- Typically worn 2 hours on and 2 hours off throughout the day
- Wrist–hand–finger splints usually are more effective because of the extension positioning of the extrinsic finger flexors.
- Rationales of efficacy of tone-reducing orthoses include:
  - Reflex-inhibiting positioning: Neurodevelopmental technique (NDT)/Bobath approach
  - Firm pressure into volar surface (palm): Rood (sensorimotor) approach
- Dorsal-based splints (e.g., Snook): Facilitation of muscle contraction by direct contact. It is theorized that stimulation of extensor surface might produce extensor muscle contraction and balance muscle tone and/or avoid increased flexor tone.
  - Functions: Flexor tone reduction, prevent skin breakdown/maceration of palm by fingernails, increase passive ROM via low-load, prolonged stretch (serial static splinting)
  - *Indications*: Spasticity due to upper motor neuron lesions (e.g., stroke, head injury, MS, cerebral palsy)

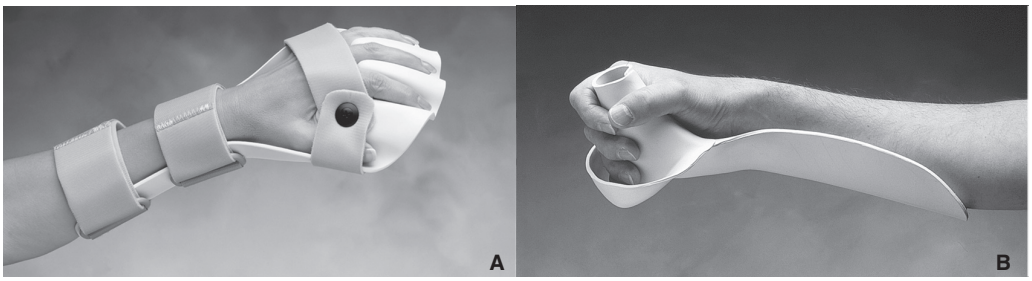


FIGURE 6–29 (A) Antispasticity "ball" splint. (B) Hand-cone splint.
Source: Courtesy of North Coast Medical, INC., Morgan Hill, CA.

SPINAL ORTHOSES

CERVICOTHORACIC ORTHOSES/(CTO)/CERVICAL ORTHOSES

Soft Cervical Collar (Figure 6–30A)

- Made of polyethylene foam or sponge rubber
- Provides no significant control of motion of the cervical spine but does provide a kinesthetic reminder (through sensory feedback) to limit motion. Retains body heat, which may help reduce muscle spasm and aid in healing of soft tissue injuries.
  - May provide comfort in part to the previously mentioned two reasons
- *Indications*: Symptomatic treatment of acute soft tissue injuries of the neck (e.g., whiplash injury)
- Maximum amount of time it should be worn: 10 days
- Risks with prolonged use: Muscle atrophy, psychological dependency

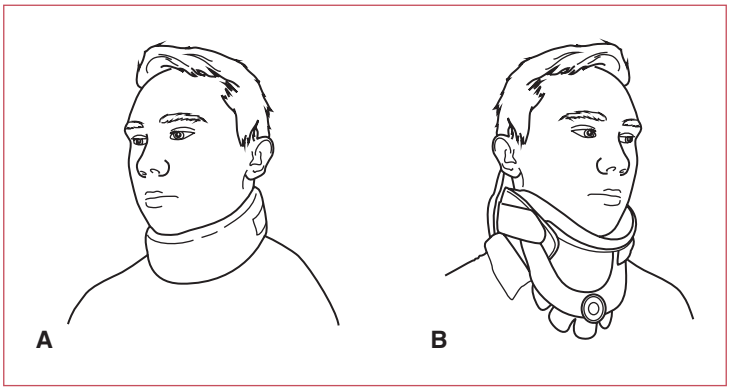


FIGURE 6–30 (A) Soft cervical collar. (B) Rigid (Philadelphia) cervical collar.

Rigid Cervical Collars

- Prefabricated orthoses that provide more restriction to cervical flexion, extension, rotation, and lateral bending than the soft collar
  1. **Thomas collar:**
     - Made of firm plastic with superior and inferior padding that wraps around the neck and is secured with Velcro
     - *Indications*: Soft tissue injuries
  2. **Philadelphia collar (Figure 6–30B):**
     - Provides total contact in the cervical spine and a mild degree of motion control
     - Made of Plastazote, has rigid anterior and posterior Kydex plastic reinforcements, and is secured by Velcro closures
     - It encompasses the lower jaw and the occiput and extends to the proximal thorax.
     - *Indications*: Soft tissue injuries and stable bony or ligamentous injuries. Also used when patients are weaned off more restrictive orthoses to limit sudden strain on the neck after prolonged immobilization.
  3. **Miami–J, Newport, and Malibu collars (variants of the Philadelphia collar):**
     - Provide better control
     - More expensive

Sterno-Occipital Mandibular Immobilizer (SOMI) (Figure 6–31)

- Cervicothoracic orthosis (CTO) with chest piece connected by uprights (going from anterior to posterior) to occipital plate. Therefore, it can easily be applied to a supine patient.

- Has removable mandibular piece so patient can eat, wash, or shave while lying supine
- *Indications*: Cervical arthritis, postsurgical fusions, stable cervical fractures

Poster-Type Cervicothoracic Orthoses

- Two- or four-poster orthoses (**Figure 6–32A, B**)
- Provide cervical spine control through mandibular and occipital components connected to sternal and thoracic components by two or four (sometimes three) posts
- Provide good control of flexion/extension; lateral bending and rotation are not well controlled.
- Can hold the head in extension or flexion by adjusting the length of anterior or posterior posters
- Cooler than cervical collars but bulkier
- *Indications*: Mid or low (with extension) stable cervical fractures and arthritis

Yale Cervicothoracic Orthoses

- Similar to an extended Philadelphia collar reinforced with rigid plastic struts extending down onto the anterior and posterior thorax with strapping beneath the axilla
- The occipital piece can extend higher than the original Philadelphia collar.

FIGURE 6–31 Sterno-occipital mandibular immobilization (SOMI) orthosis.
Source: Redrawn from Braddom RL. *Physical Medicine and Rehabilitation*. W. B. Saunders; 1996:334–336, 1194–1196, with permission.

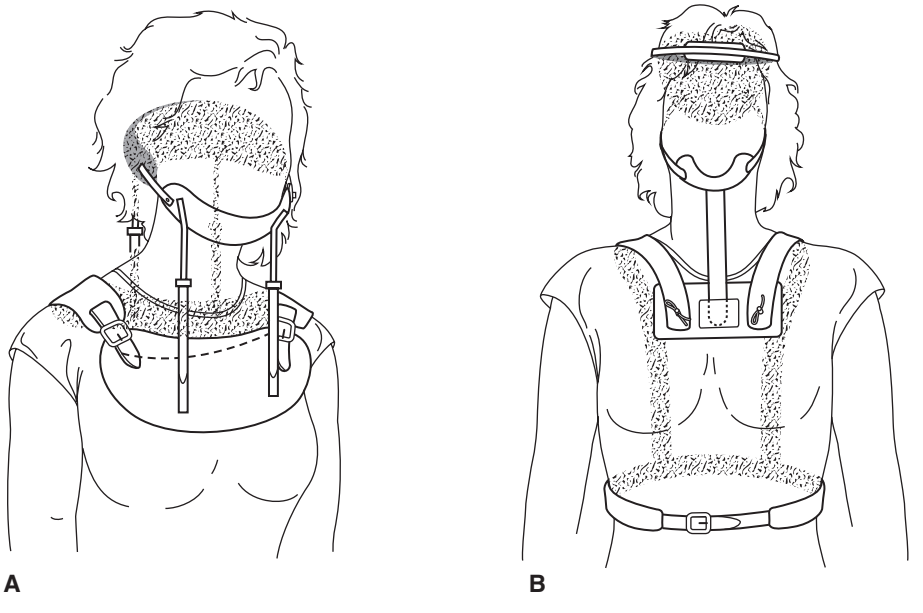


FIGURE 6–32 (A) Four-poster collar. (B) Two-poster orthosis.
Source: Redrawn from Braddom RL. *Physical Medicine and Rehabilitation*. W. B. Saunders; 1996:334–336, 1194–1196, with permission.

🕮 Minerva Cervicothoracic Orthosis/Thermoplastic Minerva Body Jacket (Figure 6–33)

- The Minerva CTO encloses the entire posterior skull, includes a band around the forehead, and extends downward to the inferior costal margin.
- Forehead band provides good control of all cervical motions.
- *Advantages*: Lighter weight than the halo vest; no pins (no "invasive" supports that carry risks of infection and slippage)
- *Disadvantages*: Less restriction of motion compared with a halo vest
- *Indications*: Management of unstable cervical spine (although halo vest use is usually preferred for maximum motion control)
  - May be the preferred orthosis (over halo) in the management of cervical spine instability in preschool-age children due to ↑ comfort, ↓ weight, and because it allows early mobilization of the patient for rehabilitation, in addition to providing the necessary stability

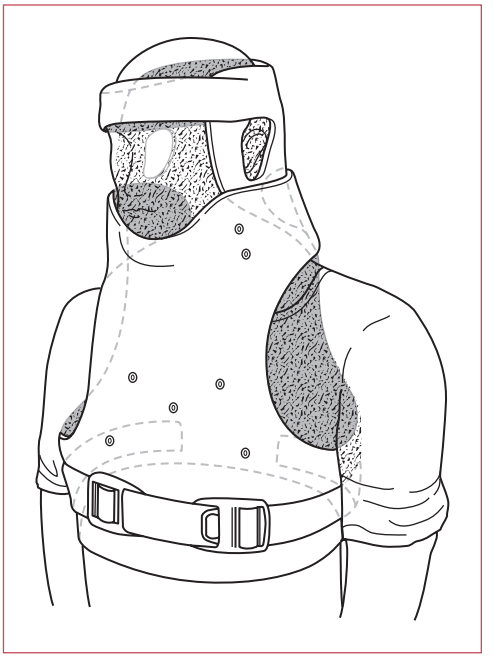


FIGURE 6–33 Thermoplastic Minerva body jacket. *Source:* Redrawn from Braddom RL. *Physical Medicine and Rehabilitation*. W. B. Saunders; 1996:334–336, 1194–1196, with permission.

🕮 Halo Vest Cervicothoracic Orthosis (Figure 6–34)

- Provides the best control of motion (all planes) in the cervical spine of all the cervical CTO
- Permits early mobilization without risk of compromising spinal alignment
- Nonremovable CTO
- Consists of a rigid halo ring secured to the skull with four external fixation pins (anterolateral above the orbital rim and posterolateral below the largest diameter of the skull—to prevent cephalad pin migration, piercing the temporalis muscle, frontal and temporal fossa, or injury to cranial nerves)
- The halo supports four posts attached to the anterior and posterior part of the vest (thoracic component).
- Of note, there has been a decline in use of halo vests due to advances in cervical surgical fusion techniques and complications from its use (Koutsogiannis et al., 2024).
- *Indications*: Management of unstable fractures of the cervical spine (especially high cervical fractures); nonsurgical alternative in cases where surgery is contraindicated or refused
- *Complications*: Risk of pressure ulcers with bedrest (usually in the region of scapula and sternum; see **Table 6–16**)

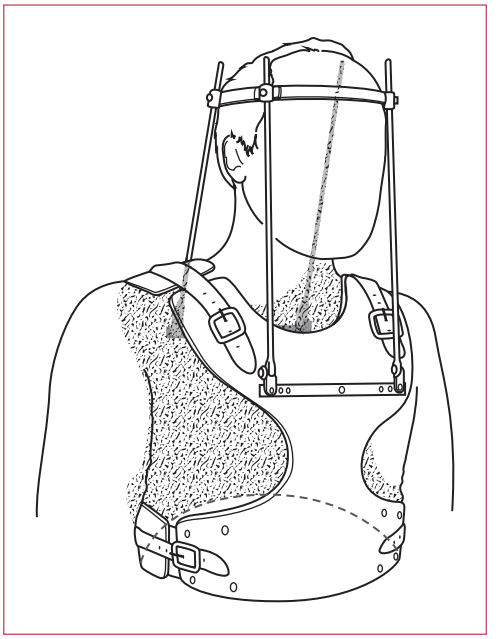


FIGURE 6–34 Halo vest. *Source:* Redrawn from Braddom RL. *Physical Medicine and Rehabilitation*. W. B. Saunders; 1996:334–336, 1194–1196, with permission.

TABLE 6–16 Complications Associated With Halo Vest Use

- Pin loosening (the most frequent complication)
- Loss of spinal reduction while wearing a halo
- Failure to develop spinal stability after wearing the halo
- Pressure sore development
- Pin site pain
- Pin tract infection
- Brain abscess
- Local osteoporosis
- Acute equilibrium impairments
- Forehead scarring
- Dysphasia
- Pin penetration of the skull
- Halo ring migration
- Cranial nerve palsy
- Brachial plexopathy
- Reduction in vital capacity

Source: From DeLisa JA, Gans BA, Walsh NE, eds. *Physical Medicine & Rehabilitation Medicine: Principles and Practice*. 4th ed. Lippincott Williams & Wilkins; 2005, with permission.

Effect of Cervical Orthoses on Normal Cervical Motion (Table 6-17)

TABLE 6–17 Normal Cervical Motion From Occiput to First Thoracic Vertebra and the Effect of Cervical Orthoses

	MEAN OF NORMAL MOTION (%)		
CERVICAL ORTHOSIS	FLEXION/ EXTENSION	LATERAL BENDING	ROTATION
(Normal motion)	100.0	100.0	100.0
Soft collar (Johnson et al., 1977)	74.2	92.3	82.6
Philadelphia collar (Johnson et al., 1977)	28.9	66.4	43.7
SOMI brace (Johnson et al., 1977)	27.7	65.6	33.6
Four-poster brace (Johnson et al., 1977)	20.6	45.9	27.1
Yale cervicothoracic brace (Johnson et al., 1977)	12.8	50.5	18.2
Halo device (Johnson et al., 1977)	4.0	4.0	1.0
Halo device (Lysell, 1969)	11.7	8.4	2.4
Minerva body jacket (Braddom, 1996; Maiman et al., 1989)	14.0	15.5	0

SOMI, sterno-occipital mandibular immobilizer.

THORACOLUMBOSACRAL ORTHOSIS

- 🕮 In general, thoracolumbosacral orthoses (TLSOs) extend from the sacrum to above the inferior angle of the scapulae and are used to support and stabilize the trunk (e.g., truncal paralysis, postspinal fusion surgery) and to prevent progression of moderate scoliosis (20–45 degrees) until patient reaches skeletal maturity, and used for thoracic kyphosis.
- Except for the thoracolumbosacral (TLS) flexion-control orthoses, TLSOs can increase intra-abdominal pressure (which in turn ↓ load on spine/intervertebral discs by transmission of the load to the surrounding soft tissues).
- They also cause an increase in O_2 consumption and energy expenditure.

- During ambulation, with axial rotation between the pelvis and the shoulders, there may be increased motion at the unrestrained segments cephalad (rostral) and caudal to the orthosis, in addition to increased energy consumption of ambulation.

Taylor Brace (Figure 6–35)

- Flexion/extension control TLSO that consists of two posterior paraspinal bars attached inferiorly to a pelvic band. An interscapular band stabilizes the paraspinal bars and serves as attachment for the axillary straps.
- The orthosis also includes a corset or anterior, full-front abdominal support, which increases intracavitary pressure.

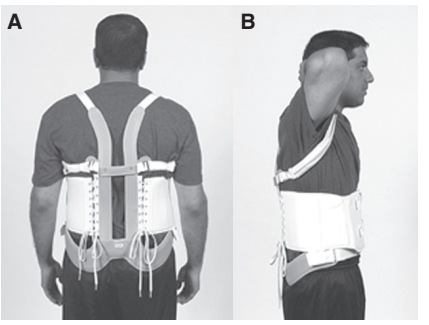


FIGURE 6–35 Taylor brace: TLSO flexion-extension control orthosis. (A) Posterior view. (B) Lateral view. TLSO, thoracolumbosacral orthosis. *Source:* Courtesy of JFK Johnson Rehabilitation Institute, 2000.

Knight-Taylor Brace (Figure 6–36)

- Consists of a Taylor-style TLSO with lateral bands and a thoracic band to restrict lateral bending
- *Indications*: Postsurgical or nonsurgical management of stable thoracic or lumbar fracture

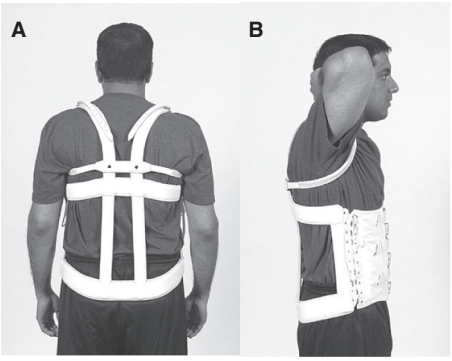


FIGURE 6–36 Knight-Taylor brace. (A) Posterior view. (B) Lateral view. *Source:* Courtesy of JFK Johnson Rehabilitation Institute, 2000.

Spine Flexion-Control Thoracolumbosacral Orthoses

🕮 JEWETT BRACE (FIGURE 6–37)

- Flexion-control TLSO consisting of a sternal pad, suprapubic pad, and anterolateral pads connected by oblique lateral uprights counteracted by a dorsolumbar pad
- The suprapubic band may be substituted by a boomerang band, which applies force on the iliac crests (used in females to avoid direct pressure on the bladder).
- *Indications:*
  - 🕮 Used to permit the upright position while preventing flexion after compression fracture of the thoracolumbar (TL) spine
    - Use in the treatment of compression fractures in osteoporotic elderly patients is controversial because it can place excessive hyperextension forces on the lower lumbar vertebrae, which can induce posterior element fractures or exacerbate a degenerative arthritis condition.
  - Thoracolumbar Scheuermann's disease
  - Used in thoracic osteoporotic kyphosis (with limited efficacy)

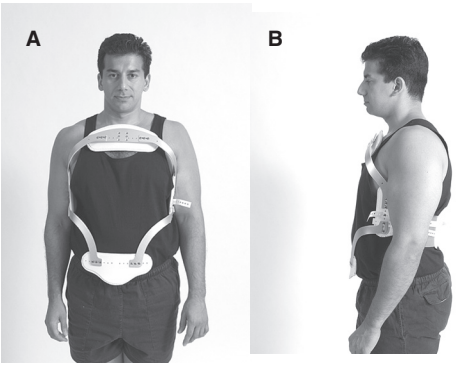


FIGURE 6–37 Jewett hyperextension orthosis. (A) Anterior view. (B) Lateral view. *Source:* Courtesy of JFK Johnson Rehabilitation Institute, 2000.

CRUCIFORM ANTERIOR SPINAL HYPEREXTENSION THORACOLUMBOSACRAL ORTHOSIS BRACE (FIGURE 6–38)

- Anteriorly, it has cross-shaped vertical and horizontal metal uprights with sternal, pubic, posterior, and anterolateral pads. The vertical upright joins the sternal and pubic pad. The horizontal uprights connect the posterior TL pad and the anterolateral pads.
- *Indications*: Similar to Jewett

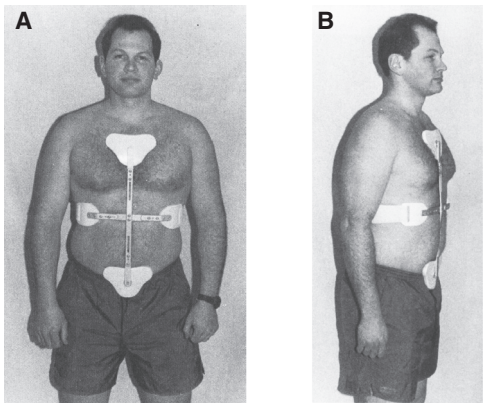


FIGURE 6–38 CASH TLSO. (A) Anterior view. (B) Lateral view.
CASH, cruciform anterior spinal hyperextension orthosis; TLSO, thoracolumbosacral orthosis.

CERVICAL–THORACIC–LUMBAR–SACRAL ORTHOSIS

Milwaukee Brace (Figure 6–39)

- 🕮 Cervical–thoracic–lumbar–sacral orthosis (CTLSO) used for scoliosis
- Consists of a rigid plastic pelvic girdle connected to a neck ring over the upper thorax by one anterior, broad aluminum bar and two posterior paraspinal bars
- The cervical ring has mandibular and occipital bars, which rest 20 to 30 mm inferior to the occiput and mandible.
- Pads strapped to the bars apply a transverse load to the ribs and spine to correct scoliotic curvatures.
- *Indications:*
  - Idiopathic or flexible congenital scoliosis with curves 25 to 40 degrees; if the curve apex is located superior to T8, the scoliosis shows signs of progression, and puberty has not finished.
  - Thoracic kyphosis in Scheuermann's disease

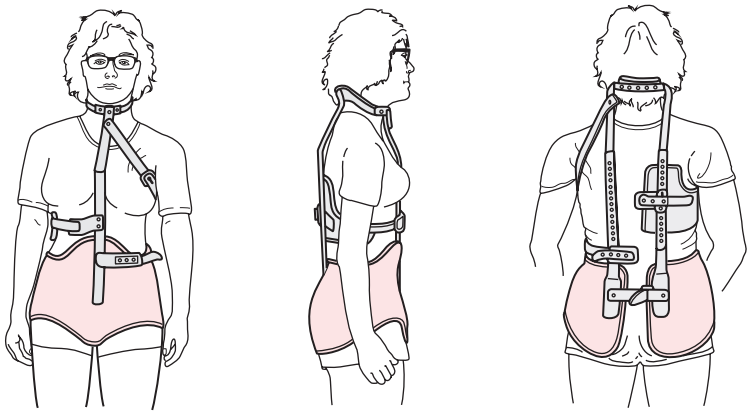


FIGURE 6–39 Milwaukee-style CTLSO. (A) Anterior view. (B) Lateral view. (C) Posterior view.
CTLSO, cervical–thoracic–lumbar–sacral orthosis.

CORSETS/FLEXIBLE SPINAL ORTHOSES

- Made of fabric/canvas with pouches for vertical stays
- A number of different types of corsets are available, including lumbosacral (LS), TLS, TL, sacroiliac, and lumbar.

- They are the most frequently prescribed orthotic for low back pain.
- The efficacy of corsets remains controversial.

Lumbar and Lumbosacral Corsets

- The most commonly prescribed lumbar-sacral support is the LS corset.
- Surrounds the torso and hips, and borders the xiphoid or lower ribs, pubic symphysis, inferior angle of the scapula, and gluteal fold. Special designs used for pregnancy, abdominal ptosis, and pendulous abdomen.
- *Indications:* Low back pain, muscle strain
- *Pros:* Kinesthetic reminder, support abdomen, reduce load on LS spine, reduce excessive lumbar lordosis, and decrease lateral bending (29%)
- *Cons:* May result in weakening of the muscles that support the trunk

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SPINAL CORD INJURIES

7

Steven Kirshblum, MD • Jeremiah Nieves, MD • Priscila Gonzalez, MD • Sara J. Cuccurullo, MD • Jennifer Chui, MD • Phillip Gordon, MD

EPIDEMIOLOGY

- 🕮 **Incidence in the United States (**National Spinal Cord Injury Statistical Center, 2023**):** 54 new injuries per million population per year—18,000 new spinal cord injury (SCI) cases per year. This number does not include those who die at the location of the incident that caused the SCI.
- 🕮 **Prevalence:** Approximately 302,000 cases
- **Gender**: Approximately 4:1 male-to-female ratio
- **Race**: 55.5% in non-Hispanic Whites; 25% non-Hispanic Blacks; 14.5% Hispanic origin; 2.6% Asian; 0.5% Native American; 1.9% other
- 🕮 **Average age at injury (since 2015):** 43 years of age
  - Persons older than 60 years of age account for >11% of SCI cases
  - Trends: ↑ in average age
  - SCIs occur most frequently among persons in the 16- to 30-year age group.
  - Children 15 years of age or younger account for only 4.5% of SCI cases.
- **Etiology:**
  - 37.6% motor vehicle crashes (MVCs)
  - 31.5% falls
  - 15.4% violence (most are gunshot wounds)
  - 8.3% sports (most common is diving)
  - Trends: ↓ in SCI due to fall-related and MVC-related SCI and an ↑ in violence
  - Falls are the most common cause of SCI in persons over 45 years old.
- **Most common time of injury:**
  - Summer season (highest incidence in July)
  - Weekends (Saturday > Sunday)
  - Time: Nighttime
- 🕮 **Type of injury:** Over the last 20 years, cases of incomplete tetraplegia have increased, while complete paraplegia and complete tetraplegia have decreased:
  - C4 is overall the most common level of injury (National Spinal Cord Injury Statistical Center, 2023)
  - T12 is the most common level of injury of paraplegia
  - Bimodal distribution of cervical spine (C-spine) fractures: 15 to 54 years and 65 to 80 years

  - 47.1% incomplete tetraplegia
  - 20.1% incomplete paraplegia
  - 19.8% complete paraplegia
  - 12.5% complete tetraplegia
- **Lengths of stay (LOS):** LOS in the hospital acute care unit has declined from 24 days in the 1970s to 12 days since 2015. Rehabilitation LOS have also declined from 98 days in the 1970s to 31 days since 2015.
- **Social and vocational demographics:**
  - Traumatic SCI patients are more likely to be single (45.1%; i.e., never married, separated, or divorced).
  - Likelihood of marriage remaining intact is lower when compared with uninjured population, both before and after injury.
  - Likelihood of marriage after SCI is also reduced.
  - 🕮 Postinjury marriages survive better than preinjury marriages.
  - 🕮 Return to work postinjury: At 1-year postinjury, approximately 18% of persons with traumatic SCI (tSCI) are employed. The employment rate increases over time to 31% at 30 years postinjury.
    - The higher the level and more severe the injury, the less likely to return to employment.
    - Education is the most important factor in being able to return to work:
      - Other predictors of returning to work include younger age, male, White, married, and ability to drive.
- **Life expectancy:**
  - Mortality rates are higher during the first year after injury than during subsequent years.
  - Overall life expectancy has somewhat improved from 50 years ago but remains below normal.
  - Mortality in the first year postinjury has greatly improved; less so for subsequent years postinjury in the last decades.
  - Predictors of mortality after injury include:
    - Male gender
    - Advanced age
    - Ventilator-dependent
    - Injured by an act of violence
    - High injury level (particularly C4 or above)
    - Neurologically complete injury
    - Poor community integration
    - Poor economic status indicators
  - The distinction between injury grades (neurologically complete vs. incomplete) is more important for those with the highest levels of injury but not for those with lower injuries.
- 🕮 **Causes of death:**
  - Respiratory disorders are the leading cause of death following SCI, both in the acute and chronic time periods, with pneumonia being the most common cause (DeVivo et al., 2022).
  - Pulmonary embolus is most common during the first year after SCI (DeVivo et al., 2022).
  - Heart disease ranks second during the chronic phase, followed by septicemia (usually associated with pressure injuries, urinary tract, or respiratory infections) and cancer.
  - Genitourinary diseases (e.g., renal failure) were the leading cause of death >40 years ago, but this has declined dramatically, most likely due to advances in urologic management.
  - Suicide risk: The suicide rate is approximately 3× that for persons of comparable age, sex, and race in the general population.
    - Suicide risk is highest during the first 5 years after SCI; for persons with paraplegia American Spinal Injury Association (ASIA) Impairment Scale (AIS) A, B, or C injury; and for non-Hispanic Whites.

ANATOMY OF THE SPINE (FIGURE 7–1)

- The spinal column consists of 33 vertebrae:
  - 7 cervical vertebrae
  - 12 thoracic vertebrae
  - 5 lumbar vertebrae
  - 5 sacral vertebrae
  - 4 coccygeal vertebrae
- 🕮 Spinal cord anatomy:
  - Located in the upper two-thirds of the vertebral column
  - The terminal portion of the cord is the **conus medullaris,** which then becomes the **cauda equina** ("horse's tail") at the L1–L2 vertebral levels
  - The spinal cord has white matter surrounding an inner core of gray matter. The white matter consists of nerve fibers, neuroglia, and blood vessels. The nerve fibers form spinal tracts, which are divided into ascending, descending, and intersegmental tracts. The location and function of various tracts are shown in Figure 7–2.

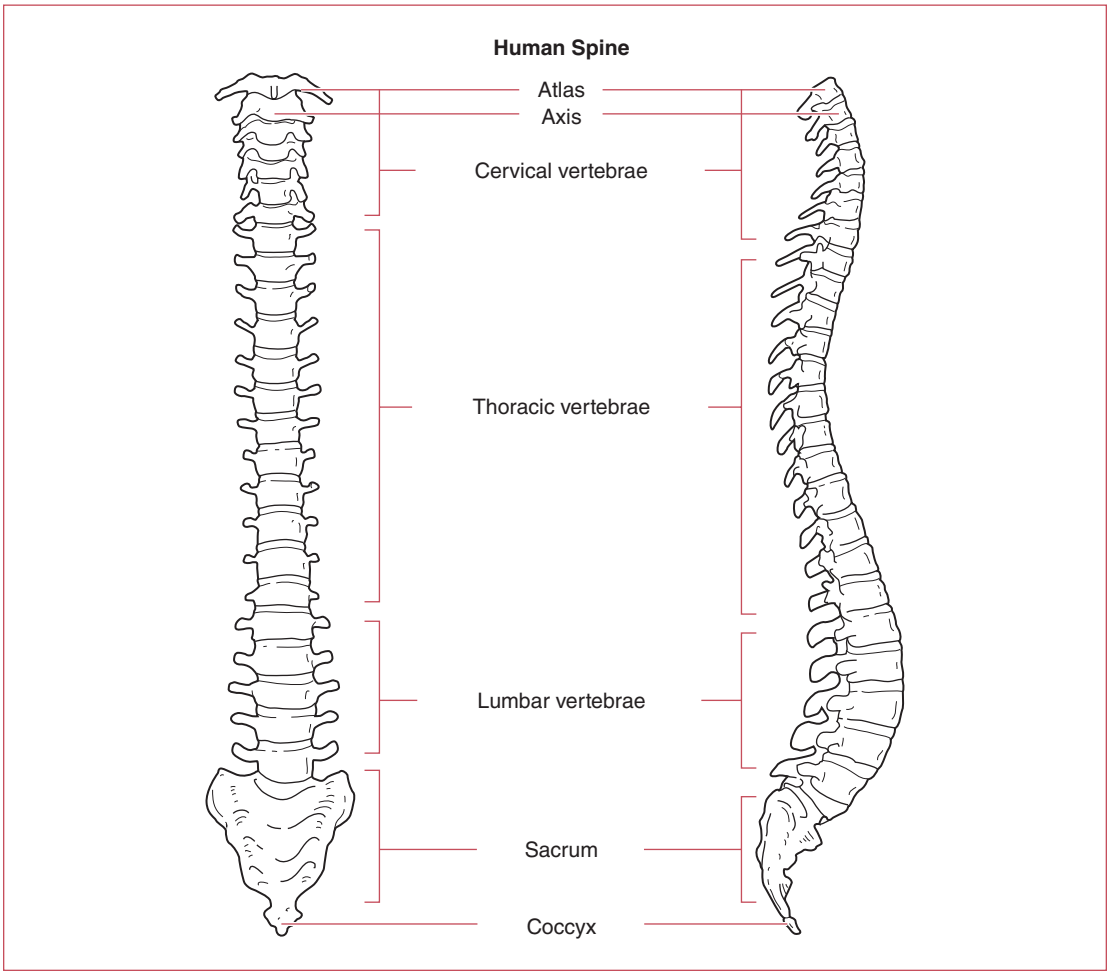


FIGURE 7–1 Human vertebral column.

LONG TRACTS IN THE SPINAL CORD			
KEY	TRACT	LOCATION	FUNCTION
	Fasciculus gracilis: Medial dorsal columns	Proprioception from the leg	Light touch Vibration
	Fasciculus cuneatus: Lateral dorsal columns	Proprioception from the arm	Light touch Vibration
	Spinocerebellar	Superficial lateral column	Muscular position and tone, unconscious proprioception
	Lateral spinothalamic	Ventrolateral column	Pain and thermal sensation
	Ventral spinothalamic	Ventral column	Tactile sensation of crude touch and pressure
	Lateral corticospinal tract (pyramidal)	Deep lateral column	Motor neuron distribution: Motor fibers theorized to run medial (cervical) → lateral (sacral) C → S
	Anterior corticospinal tract	Medial ventral column	Motor: Neck and trunk movements

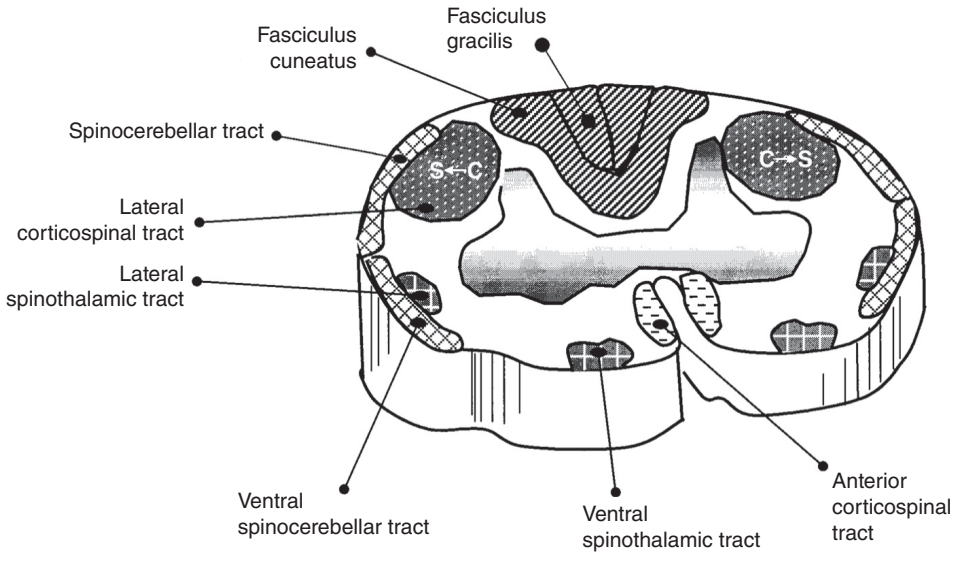


FIGURE 7–2 Transverse section of the spinal cord (use the key for long tracts, location, and function).

MAJOR ASCENDING AND DESCENDING PATHWAYS IN THE SPINAL CORD (FIGURE 7–3)

Note where tracts cross in relation to the brainstem in Figure 7–3.

Descending Pathways

- **Lateral corticospinal tracts:**
  - Main motor tracts for controlling voluntary muscle activity
  - Its origin is in the precentral gyrus of the frontal lobe of the brain. Axons descend through the internal capsule to the medulla oblongata.
  - 80% to 90% of the axons cross over (decussate) to the contralateral side at the pyramidal decussation in the medulla.
  - Nerve fibers then descend in the lateral white columns of the spinal cord (lateral corticospinal tracts). At each level of the spinal cord, the axons from the lateral tract peel off and enter the gray matter of the ventral horn to synapse with secondary neurons.
  - The remaining 10% to 20% of axons that do not decussate/travel in the anterior (ventral) corticospinal tracts. The axons of the ventral tract then cross over at the corresponding level of muscles that they innervate.
  - Both tracts travel from the precentral gyrus to the ventral horn as uninterrupted neurons and are termed upper motor neurons (UMNs), while the secondary neurons that they synapse on are termed lower motor neurons (LMNs).
  - Cerebral lesions result in contralateral deficits in general.

Ascending Pathways

- **Spinocerebellar tracts:**
  - Transmit unconscious proprioception (muscle proprioceptive, stretch, tension fibers) from the **ipsilateral** side of the body to the brain
  - Because these tracts remain ipsilateral, cerebellar lesions produce ipsilateral malfunctioning.
- **Lateral spinothalamic tracts:**
  - Transmit pain and temperature from the **contralateral** side of the body to the brain
  - Pain and temperature sensory fibers enter the spinal cord and synapse in the dorsal horn of the gray matter. The fibers cross over to the contralateral side of the spinal cord within one to three vertebral segments, ascend in the lateral spinothalamic tracts to the contralateral thalamus, and then ascend in the internal capsule to the postcentral gyrus of the cerebral cortex.

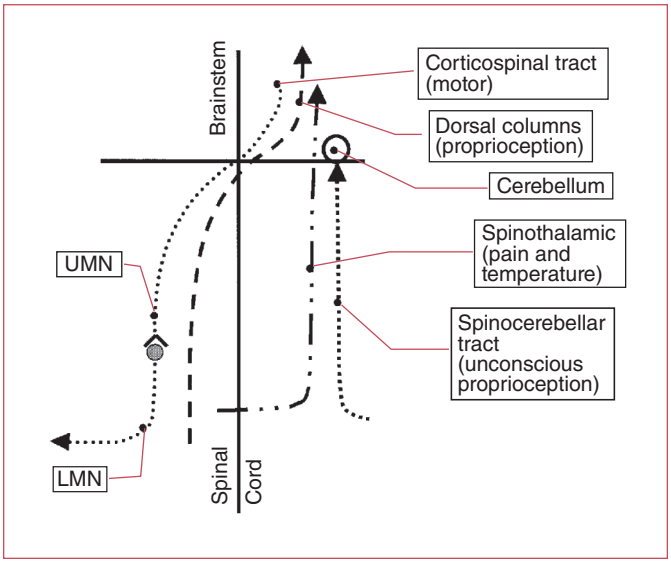


FIGURE 7–3 A schematic view: The major long tracts in the spinal cord (ascending and descending arrows depict direction). LMN, lower motor neurons; UMN, upper motor neurons.

  - A lesion of the lateral spinothalamic tract will result in loss of pain-temperature sensation contralaterally below the level of the lesion.
- **Dorsal (posterior) columns:**
  - Transmit proprioception, fine touch, and vibration sense from the **ipsilateral** side of the body to the brain
  - These sensory fibers synapse at the dorsal root ganglion and immediately ascend into the ipsilateral dorsal white columns.
  - They travel up to the medulla, at which point they decussate. Axons that enter the cord at the sacral and lumbar levels are situated in the medial part of the dorsal column (i.e., the lower part of the body), called the fasciculus gracilis. Those axons that enter at the thoracic and cervical levels are situated in the lateral part of the column (from the upper part of the body) and are termed the fasciculus cuneatus. Axons of each fasciculus synapse in the medulla and form a bundle termed the *medial lemniscus*, which ascends to the postcentral gyrus.
  - A lesion of the posterior columns results in the loss of proprioception and vibration ipsilaterally below the level of the lesion.

Blood Supply of the Spinal Cord (Figure 7–4)

- The spinal cord receives blood supply from one anterior and two posterior spinal arteries as well as anterior and posterior radicular arteries.
- The anterior spinal artery arises as a single artery that runs within the anterior median fissure and supplies blood flow to the anterior two-thirds of the spinal cord.
- Posterior spinal arteries arise directly or indirectly from the vertebral arteries, run inferiorly along the sides of the spinal cord, and provide blood to the posterior one-third of the spinal cord.
- Radicular arteries are branches of local arteries (vertebral, cervical, intercostals, lumbar, and sacral) that enter the vertebral canal through the intervertebral foramina and reinforce the anterior and posterior spinal arteries.
  - 🕮 The **artery of Adamkiewicz** provides the major blood supply to the lumbar and sacral cord. It generally arises from the left intercostal or lumbar artery at the levels of T9–L3 and provides the major blood supply to the lower two-thirds of the spinal cord.
  - 🕮 The mid-lower thoracic region is referred to as the "watershed area" because there are fewer radicular arteries that supply the mid-thoracic region of the spinal cord. This area (T4–T6) is most affected when there is low blood flow to the spinal cord (i.e., clamping of the aorta in surgery).
  - The veins of the spinal cord drain mainly into the internal venous plexus.

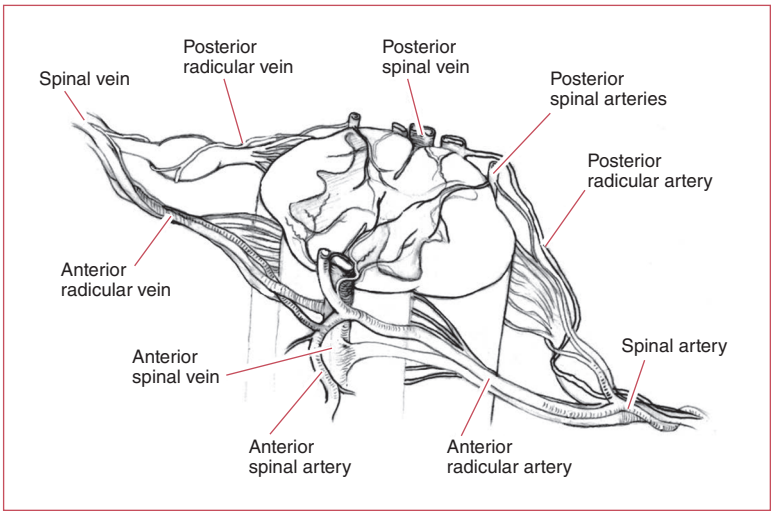


FIGURE 7–4 Arterial and venous supply to the spinal cord (transverse section).

SPINAL PATHOLOGY

CERVICAL SPINE FLEXION/HYPEREXTENSION INJURIES (SEE TABLE 7–1)

TABLE 7–1 Cervical Flexion/Extension Injuries

MECHANISM OF INJURY	STABILITY	POTENTIAL INJURY	MOST COMMON LEVEL
Flexion/axial loading (i.e., diving) *Burst/compression fracture*	Stable if ligaments remain intact	Compression fracture with fragmentation of vertebral body and projection of bony spicules into canal	C5
Flexion/rotation injury *Unilateral facet dislocation*	Unstable if PLL disrupted Vertebral body <50% displaced on x-ray	Likely to be incomplete SCI if spinal cord is compromised	C5–C6
Flexion *Bilateral facet dislocation*	Unstable if PLL disrupted Vertebral body displaced >50% on x-ray	Ant. dislocation of C-spine with spinal cord compression/compromise More likely to result in complete SCI	C5–C6
Hyperextension *Central cord syndrome*	Stable; anterior longitudinal ligament may be disrupted	Hyperextension of C-spine Clinically: UE weaker than LE Is an incomplete injury	C4–C5

C-spine, cervical spine; LE, lower extremity; PLL, posterior longitudinal ligament; SCI, spinal cord injuries; UE, upper extremity.

Flexion Injuries

BURST/COMPRESSION FRACTURES (FIGURE 7–5)

- Mechanism: Cervical flexion with axial loading
- C5 is the most common compression fracture of the C-spine.
- Force ruptures the plates of the vertebra and compresses the body. Anterior wedge-shaped-appearing vertebrae are typically seen on x-ray.
- Fragments may project into the spinal canal, which may result in injury to the nerve root and/or cord itself with retropulsion of bony fragments.

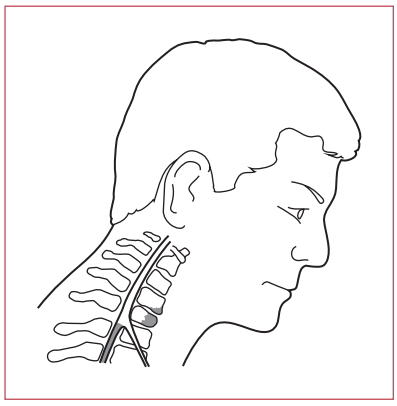


FIGURE 7–5 Cervical burst/compression fracture.

UNILATERAL FACET JOINT DISLOCATIONS (FIGURE 7–6)

- Mechanism: Cervical flexion–rotation injury
- Vertebral body <50% displaced on x-ray
- Unstable if the posterior longitudinal ligament (PLL) is disrupted
- Narrowing of the spinal canal and neural foramen
- C5–C6 most common level
- Also note that flexion and rotation injuries may disrupt the intervertebral disc, facet joints, and interspinous ligaments with little or no fracture of the vertebrae.
- If SCI results, it is more likely to be an incomplete injury.

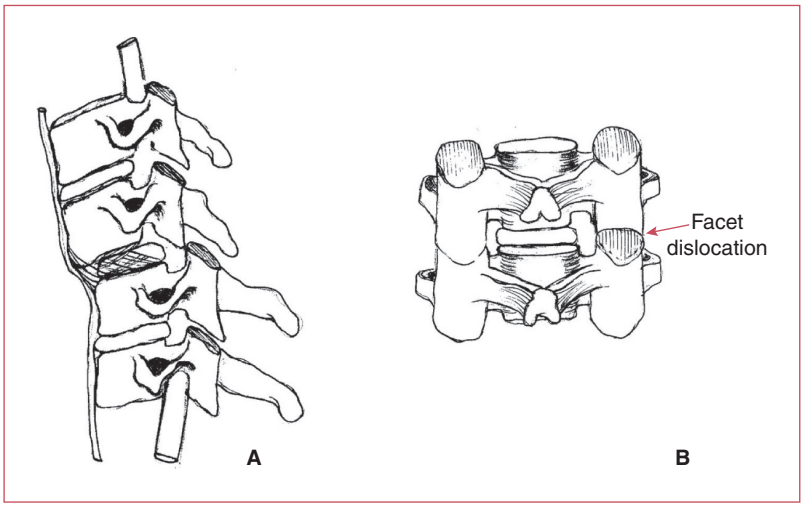


FIGURE 7–6 Unilateral facet joint dislocation. (A) Lateral view. Note: There is <50% anterior dislocation of the vertebral body. (B) Posterior view.

BILATERAL FACET JOINT DISLOCATIONS (FIGURE 7–7)

- Mechanism: Flexion injury
- Vertebral body >50% displaced on x-ray, causing significant narrowing of the spinal canal
- Unstable with disruption of the PLL
- Most common level is C5–C6 because of increased movement in this area.
- Injury is more likely to be neurologically complete.

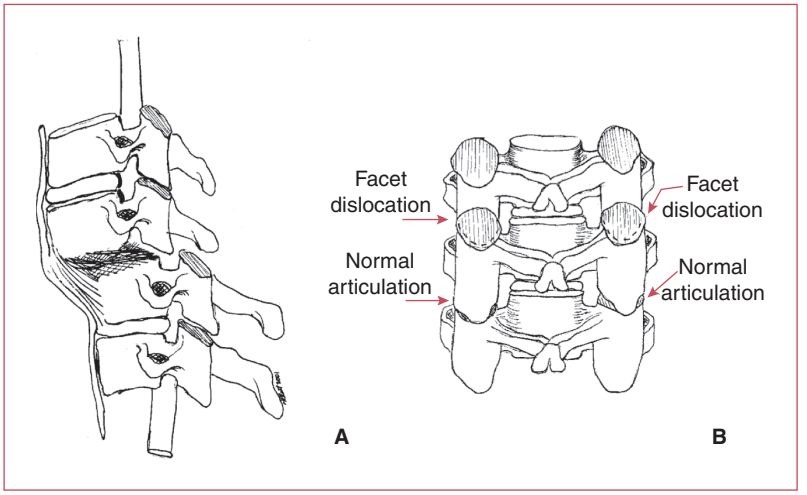


FIGURE 7–7 Bilateral facet joint dislocation. (A) Lateral view. Note: There is >50% anterior dislocation of the vertebral body. (B) Posterior view.

Hyperextension Injuries (Figure 7–8)

- Can be caused by acceleration-deceleration injuries, such as MVC
- C4–C5 is the most commonly affected level.
- Soft tissue injury may not be seen on radiologic studies.
- Hyperextension injury of the C-spine in the elderly may result in a central cord syndrome (CCS)—see section on Central Cord Syndrome later in this chapter for more detail.

NONTRAUMATIC SPINAL CORD INJURY

- Nontraumatic (NT)-SCI etiologies include spinal stenosis with myelopathy, spinal cord compression from a neoplasm, multiple sclerosis (MS), transverse myelitis (TM), infection (viral, bacterial, fungal, parasitic abscess), vascular ischemia, radiation myelopathy, motor neuron diseases, syringomyelia, vitamin B12 deficiency, and others.
- Spinal stenosis with myelopathy and spinal cord tumors are the most common causes of NT-SCI presenting for inpatient rehabilitation in the United States.

FIGURE 7–8 Cervical spine hyperextension injury.

SPINAL STENOSIS WITH MYELOPATHY

- Cervical spinal stenosis due to spondylosis is the most common cause of myelopathy.
- Spondylotic changes result in disc space narrowing, facet hypertrophy, osteophytes from vertebral bodies, and ligamentum flavum hypertrophy, which can lead to stenosis of the spinal canal and cord compression.
- Spondylosis also causes stiffness of the spinal segments and results in relative hypermobility or subluxation of adjacent segments leading to myelopathy.
- In extension, the sagittal diameter of the cervical canal decreases: The cord may be pinched between the disc and the osteophytes from the anterior vertebral bodies.
- With cervical flexion, the cord may be tethered over spondylotic anterior elements.
- **Presentation**: Most common is gait disturbance and decreased balance (myelopathic gait), followed by upper extremity (UE) paresthesias, dexterity issues with decreased fine motor coordination, and arm/hand weakness. CST involvement first with resultant leg weakness. This is followed by posterior column involvement with presentation of a wide-based, ataxic gait.
- Physical exam signs:
  - Hand intrinsic musculature atrophy
  - Sensory loss—vibratory sense or proprioception in the extremities, especially in the feet
  - Hyperreflexia
  - Positive Hoffman's sign (reflex contraction of the thumb and index finger after flicking the end of the middle finger) and positive Babinski sign
  - Myelopathic gait
- MRI is the gold standard for evaluation of cervical myelopathy and helps to identify the pathologic causes.
- Treatment:
  - Asymptomatic to mild cases: Conservative treatment including therapy to improve gait pattern is recommended with close follow-up.
  - Moderate to severe myelopathy: Surgery may be indicated. Either an anterior or posterior surgical approach can be performed with benefits and risks associated with each. Anterior surgery has the advantage of being able to remove ventral osteophytes, although it has a higher early postoperative complication rate, including swallowing complications.

TRANSVERSE MYELITIS

- TM is inflammation of the spinal cord that presents with varying degrees of weakness, sensory alterations/deficits, autonomic dysfunction, and deep tendon reflex abnormalities. Most commonly affects the thoracic spinal cord.
- Symptoms evolve over several hours to several weeks, initially beginning with leg paresthesias. Pain is usually present at the involved cord segments, usually the upper thoracic area; it affects approximately one-third of patients.
- Weakness follows, usually associated initially with acute loss of reflexes but later with development of hyperreflexia.
- The female to male ratio is approximately 4:1, peaking in the second and fourth decades.

- Etiology: Causes include idiopathic, MS, infections, and autoimmune or postinfectious inflammation.
- Diagnostic testing includes MRI and lumbar puncture (cerebrospinal fluid [CSF] analysis):
  - MRI usually shows an intramedullary fusiform cord enlargement over several segments. There is a subtle to obvious hyperintense signal on T2-weighted and short T1 inversion recovery imaging.
  - CSF frequently shows lymphocytic pleocytosis and protein elevation but can be normal. Gamma globulin may be selectively elevated.
- Prognosis: Approximately one-third of patients with TM recover completely, one-third do not improve at all, and one-third improve but with significant residual neurologic deficit.
  - Rapid progression, back pain, and spinal shock predict poor prognosis.
- In 5% to 10% of cases, TM is the presenting feature of MS. These patients are likely to have asymmetric clinical findings, MR lesions extending over fewer than two spinal segments, abnormal visual evoked potentials, and CSF oligoclonal bands. Patients with more incomplete lesions tend to be at higher risk for progression to MS, whereas those with complete TM are less likely to convert to MS.

🕮 NEUROMYELITIS OPTICA

- Also termed Devic's disease, neuromyelitis optica (NMO) is a fairly uncommon, autoimmune demyelinating and axonal damaging disease of the central nervous system (CNS) that affects the optic nerves and the spinal cord, causing a combination of optic neuritis and TM.
- Often mimicking MS, the myelopathy in NMO is more severe than in MS, and on MRI the lesions tend to be more longitudinal (more than three spinal segments). These lesions can lead to weakness or complete paralysis, painful spasms, sensory loss, and bowel and bladder dysfunction. The optic neuritis can cause blindness in one or both eyes.
- Most of these lesions cause permanent deficits, although some flare-ups can be reversible.
- Marked female predominance
- Treatment for NMO includes intravenous (IV) glucocorticoids followed by plasmapheresis if not responsive to steroids. Intravenous immunoglobulin (IVIg) has also been considered.
- Long-term immunosuppression has become standard with rituximab, mycophenolate, mofetil, or azathioprine, but no controlled clinical trials exist for any of these interventions.

EPIDURAL ABSCESS

- Most commonly seen in diabetic and immunocompromised patients including immune suppression, IV drug use, bacterial endocarditis, and occasionally with genitourinary infection, and with poor dentition or dental infection
- Male predominance
- The initial symptom is insidious and axial spinal pain, usually severe and sharp. Fever is present in about two-thirds. Radicular pain and leg weakness, usually bilateral, occur in about 50%.
- Symptoms evolve over a few days to weeks and have been present for more than 2 weeks before diagnosis in more than two-thirds of cases.
- Lumbar puncture should be avoided because of the danger of increasing spinal cord or cauda equina compression and the risk of introducing infection into the subarachnoid space, thus producing meningitis.
- MRI is the optimal diagnostic procedure.
- Treatment consists of surgical drainage and excision of the abscess. Broad spectrum IV antibiotics should be started immediately, and when the bacterium has been identified (*Staphylococcus aureus* in over 50% of cases), antibiotic coverage can be narrowed.
- Even though surgery is the treatment of choice, there has been a trend toward medical management in patients who are neurologically intact, but they should be closely monitored, as many of those patients end up requiring surgical decompression (Adogwa et al., 2014; Arko et al., 2014; Patel et al., 2014).
- Relationship between the time of spinal cord decompression and outcome: Patients treated before the development of paralysis usually recover completely, whereas patients who are paraplegic or tetraplegic for over 36 to 48 hours usually do not.

RADIATION MYELOPATHY

- Delayed complication of radiation to lesions of the spine or adjacent tissues that develops months or years after treatment
- Incidence is correlated with the total radiation dose, the dose fraction, and the length of the spinal cord irradiated.
- Radiation myelopathy is overall rare after radiation; <1% who received radiation treatment develop radiation myelopathy.
- Clinically, there is weakness and loss of sensation, and sometimes a Brown-Séquard-like syndrome develops.
- Pathologically, the white matter of the cord is primarily affected with axonal loss and demyelination in the area exposed to radiation.
- The established diagnostic criteria include exclusion of other etiologies, the gradual development of sensory and motor loss below the level of radiation, and sufficient radiation dose and latency.
- There is generally a long latency between the completion of radiation and symptom onset. Symptoms appear after 6 months and most commonly begin after 9 to 15 months.
- There is no effective treatment. Fortunately, focused radiation protocols for tumor treatment have improved over time so that radiation myelitis has become less common.
- Another radiation-related syndrome, called **acute transient radiation myelopathy**, consists of a Lhermitte's phenomenon in patients who have had radiation exposure of the cervical cord. There are no associated abnormal sensory or motor findings. The onset of symptoms usually occurs about 4 months after radiation therapy and spontaneously resolve several months later.

SUBACUTE COMBINED DEGENERATION (LICHTHEIM'S DISEASE)

- Degeneration of the posterior **and** lateral columns of the spinal cord
- Most severely affected are the posterior columns at the cervical and thoracic levels.
- Usually begins in the thoracic region and becomes ascending or descending:
  - Insidious onset
  - Hand and feet paresthesias—tingling/numbness that progressively worsens. Loss of posterior column function and sensory ataxia.
  - Weakness of legs, arms, and trunk
  - Bilateral spastic paresis of lower limbs. Legs are typically more severely affected.
  - Hyperreflexia and positive Babinski sign may be seen (reflexes may be depressed if polyradiculopathy present).
- **Causes:**
  - Results from vitamin B12 deficiency (most common), vitamin E and copper deficiencies, and folate deficiency (rare).
  - Vitamin B12—deficiency, malabsorption, lack of intrinsic factor or inhibition of attachment of intrinsic factor, gastritis:
    - Most commonly associated with pernicious anemia
    - Nitrous oxide administration—enhances oxidation of vitamin B12, making it ineffective
  - Vitamin E deficiencies due to malabsorption: Cystic fibrosis and Bassen–Kornzweig syndrome
- **Diagnosis:**
  - Complete blood count (CBC): Megaloblastic anemia
  - Decreased serum vitamin B12 level
  - Methylmalonic acid and homocysteine (useful for cases with low normal B12 level—200–350 pg/mL and use of nitrous oxide)
  - In gastritis, look for anti-intrinsic factor antibodies and elevated serum gastrin levels.
  - MRI: T2 images with increased signal in posterior columns, spinothalamic tracts
- **Treatment:**
  - Vitamin B12 injection 1,000 µg daily for 1 week; then 1,000 µg weekly × 1 month; and monthly injections after

- Serum homocysteine and methylmalonic acid levels return to normal levels in 2 weeks with treatment. Neurologic improvement takes more time and may be incomplete depending on the extent and duration of neurodegeneration.

SPINAL CORD TUMORS

- Spinal neoplasms represent 15% of CNS tumors and are classified as either extradural or intradural. Intradural and extradural tumors are approximately equal in prevalence.
- The most common tumors that arise within the spinal cord or dura (intradural) are primary tumors. However, the most common tumors outside the dura mater (extradural) and secondarily compromise the spinal cord are metastatic, which are approximately 25 times more prevalent than primary neoplasms.
- Intradural neoplasms are further divided into those that are *intramedullary*, arising from the substance of the spinal cord (account for approximately one-third of the primary intradural spinal tumors); or *extramedullary* (approximately two-thirds of intradural spinal tumors), arising from elements extrinsic to the spinal cord. Nerve sheath tumors, meningiomas, and filum terminale ependymomas account for most of primary extramedullary neoplasms and are usually considered benign and amenable to complete surgical resection.
- **Intradural intramedullary tumors**:
  - Glial tumors, such as astrocytomas and ependymomas, represent up to 80% of intramedullary spinal cord tumors.
  - There is a predominance of astrocytomas in the pediatric population and a predominance of ependymomas in adults (frequency peaks between the second and fourth decades, affecting males and females equally, and most often involve the cervical or cervicothoracic region).
  - Ependymomas are usually associated with a slowly progressive presentation of back pain developing over months rather than weeks.
- **Intradural extramedullary spinal cord tumors:**
  - The most common primary extramedullary lesions are derived from nerve sheath cells (schwannomas and neurofibromas) or arachnoid cap cells (meningiomas), which affect the adult population with equal frequency. These three tumor types account for nearly 95% of all intradural extramedullary spinal cord tumors.
  - Nerve sheath tumors affect men and women equally, with peak incidence in the fifth decade.
  - Approximately 80% of meningiomas involve the thoracic spine. Myxopapillary ependymomas are extramedullary tumors arising from the conus medullaris and filum terminalis.
  - The most common presenting symptom is local or radicular pain secondary to spinal cord compression, especially with lesions involving the lumbar spine. In general, extramedullary tumors are histologically benign and amenable to complete surgical resection.
- 🕮 **Extradural spinal tumors:** Extradural tumors largely comprise spinal metastases and primary bony tumors. Extradural metastases account for up to 95% of spinal lesions.
  - Highest incidence between ages 40 and 65 years of age
  - The majority of cases involve the thoracic spine, followed by the lumbar spine and then the C-spine.
  - The most common primary malignancies with spinal metastases are lung, breast, and prostate cancer, as well as lymphoma.
  - The major presenting symptoms are nonspecific and include local or radicular pain with or without motor weakness, sensory changes, as well as loss of sphincter control. Radicular pain is usually unilateral in the cervical and lumbar regions but bilateral with thoracic invasion. The pain is characteristically worsened by movement and usually worse at night.
  - The risk for spinal cord involvement depends on the primary source and is highest for breast, prostate, and lung.
- **Primary bony spinal tumors:** Multiple myeloma and plasmacytoma are the most common primary malignant tumors of the spinal column in adults. These tumors are more prevalent among the African American population, affecting more males than females. This is followed by hemangiomas and osteoid osteomas.

NONTRAUMATIC SPINAL CORD INJURY RELATIVE TO TRAUMATIC SPINAL CORD INJURY

- There is a lower incidence of secondary medical complications including spasticity, orthostasis, deep vein thrombosis (DVT), pressure injuries, autonomic dysreflexia (AD), and wound infections during rehabilitation in patients with NT-SCI.
- Inpatient rehabilitation LOS are shorter for persons with NT-SCI, although home discharge rates are overall comparable to traumatic SCI.
- There is a favorable discharge to home for patients who have an incomplete injury, are married, have an established bowel and bladder management program, have intact skin, are male gender, and are cognitively intact.

CERVICAL BRACING

(See Chapter 6, Prosthetics and Orthotics)

REMOVABLE CERVICAL ORTHOSES		NONREMOVABLE CERVICAL ORTHOSES
Least restrictive ↓ Most restrictive	Soft collar	Halo brace is the most restrictive cervical orthosis of all cervical orthoses.
	Head–cervical orthosis	
	SOMI brace	
	Four-poster	
	Minerva brace	

SOMI, sterno-occipital mandibular immobilizer.

🕮 Cervical Bracing

- The soft cervical collar is the least restrictive of the collars.
- The head–cervical orthoses are often used post-surgery in SCI and include the Philadelphia®, Aspen®, Aspen Vista®, Miami J®, and Johnson Malibu.
- Minerva brace is the most restrictive removable brace, followed by the four-poster, the sterno-occipital mandibular immobilizer (SOMI).
- The Halo brace is the most restrictive and is not removable.
- See Chapter 6, Prosthetics and Orthotics, for a more in-depth discussion on spinal bracing.

Neurologic Injury Relative to Radiologic Findings

- 🕮 **Complete lesions** are more often seen with:
  - Bilateral cervical facet dislocations
  - Thoracolumbar flexion–rotation injuries
  - Trans-canal gunshot wounds
- 🕮 **Incomplete injuries** are more often seen with:
  - Cervical spondylosis—falls
  - Unilateral facet joint dislocations
  - Noncanal penetrating gunshot/stab injuries

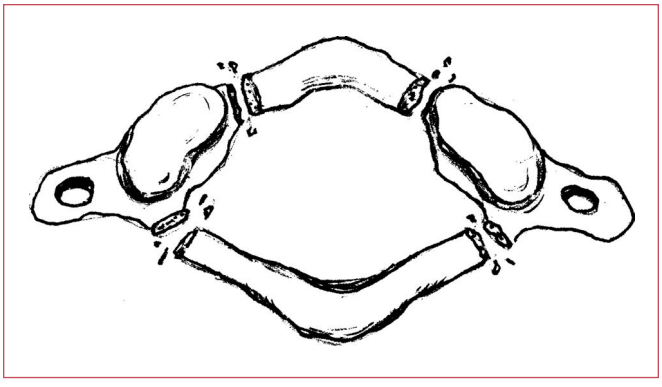


FIGURE 7–9 Jefferson fracture of C1 (superior view).

FRACTURES OF THE SPINE

Cervical Fractures

- **Jefferson fracture (C1 burst fracture; Figure 7–9):**
  - Burst fracture of the C1 ring. Usually a stable fracture with no neurologic findings.
  - Mechanism: Axial loading causing fractures of anterior and posterior parts of the atlas (i.e., football spearing)

  - Treatment: Rigid orthosis (i.e., Halo vest) if it is a stable fracture. Unstable fractures will require surgery.
- 🕮 **Hangman fracture (C2 burst fracture; Figure 7–10):**
  - Usually bilateral from an abrupt deceleration injury (e.g., MVC with head hitting the windshield)
  - Most often stable with only transient neurologic findings
  - Treatment: External orthosis (Halo is first-line treatment). Unstable fracture will require surgery (Bristol et al., 2005).
- **Odontoid (dens) fracture (Figure 7–11):**
  - Type I: Fracture through the tip of dens. No treatment usually required.
  - Type II (most common): Fracture through the base of odontoid at junction with the C2 vertebra. Can be treated with a Halo vest, surgical fusion, or cervical collar depending on age, displacement, or instability.
  - Type III: Fracture extends from base of odontoid into the body of the C2 vertebra proper. Usually treated with a Halo vest or cervical collar.
- **Vertebral body compression fracture (see Table 7–1)**

Thoracolumbar Fractures

- 🕮 **Chance fracture (Figure 7–12):**
  - Transverse fracture of the thoracic or lumbar spine from posterior to anterior through the spinous process, pedicles, and vertebral body
  - Usually affects T12, L1, and L2 levels
  - Previously was most commonly seen in patients wearing lap seat belts. Now typically due to falls/crush injury with acute hyperflexion of the thorax.
  - Tend to be stable fractures and are seldom associated with neurologic compromise unless a significant amount of translation occurs

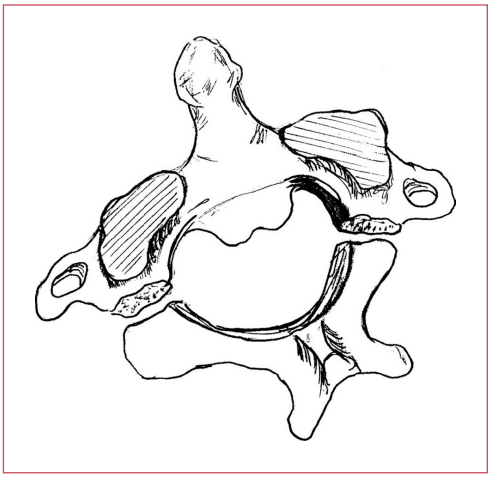


FIGURE 7–10 Hangman fracture of C2 (superior posterior view).

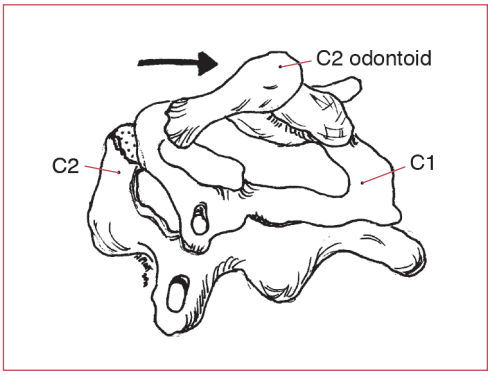


FIGURE 7–11 Odontoid fracture.

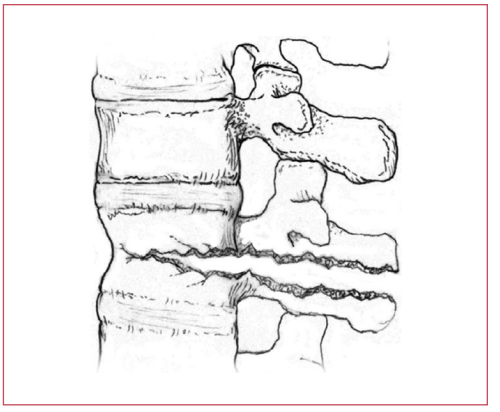


FIGURE 7–12 Chance fracture.

- **Vertebral body compression fracture (anterior wedging; Figure 7–13):**
  - Most commonly caused by axial compression with or without flexion: Anterior wedging with decreased vertebral body height—may cause thoracic kyphosis (Dowager hump)
  - Spontaneous vertebral compression fractures are stable injuries—ligaments remain intact.

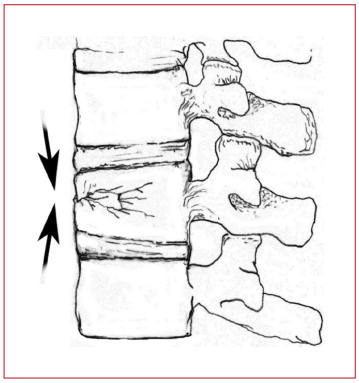


FIGURE 7–13 Vertebral body compression fracture.

SPINAL CORD INJURY WITHOUT RADIOLOGIC ABNORMALITY (SCIWORA)

- Clinical signs of SCI without evidence of fracture or malalignment on x-ray or CT
- **Children:**
  - Most commonly occurs in children
  - Predilection for the C-spine
  - **Mechanisms of injury:**
    - Traction in a breech delivery
    - Violent hyperextension or flexion
  - Predisposing factors:
    - Large head-to-neck size ratio
    - Elasticity of the fibrocartilaginous spine
    - Horizontal orientation of the planes of the cervical facet joints
- **Older adults:**
  - **Typical mechanism of injury:**
    - A fall with hyperextension of the neck, leading to an acute central cord syndrome
    - Ligamentum flavum may bulge forward into the central canal and narrow the sagittal diameter as much as 50%.
- **Imaging:**
  - Flexion/extension films should be performed cautiously only after static neck films have been cleared by a radiologist.
  - MRI may be useful to detect soft tissue pathology such as ligamentous injuries as well as cord edema or hematomas.
- **Treatment**: Typically managed nonoperatively with immobilization and therapy

SPINAL CORD INJURY CLASSIFICATION

TERMINOLOGY

- **Tetraplegia:**
  - Replaced the term *quadriplegia* in 1992
  - Impairment or loss of motor and/or sensory function in the cervical segments of the spinal cord due to damage of neural elements within the spinal canal
  - Results in impairment of function in all four extremities as well as the trunk and pelvic organs
  - Does not refer to peripheral nerve or brachial plexus injuries (i.e., refers only to UMN injuries)
- **Paraplegia:**
  - Impairment or loss of motor and/or sensory function in the thoracic, lumbar, or sacral (but not cervical) segments of the spinal cord
  - Trunk, legs, and pelvic organs may be involved, but arm function is spared.
  - Also refers to cauda equina and conus medullaris injuries but not to LMN injuries outside the neural canal, such as lumbosacral plexus lesions or peripheral nerve injury
- **Dermatome:**
  - Area of skin innervated by the sensory axons within each segmental nerve (root)
- **Myotome:**
  - Collection of muscle fibers innervated by the motor axons within each segmental nerve (root)

UPPER MOTOR NEURON VERSUS LOWER MOTOR NEURON INJURY

UMN INJURY	LMN INJURY
Innervation: Begins in the prefrontal motor cortex, travels through the internal capsule and brainstem, and projects into the spinal cord	Innervation: Begins with the anterior horn cells of the spinal cord and includes the peripheral nerves
UMN INJURY FINDINGS	**LMN INJURY FINDINGS**
Hyperreflexia Pathologic reflexes (e.g., Babinski response) Detrusor sphincter dyssynergia (depending on level of lesions)	Hyporeflexia Flaccid weakness Significant muscle atrophy Areflexic/hypotonic bladder

Note: Lesions of the upper lumbar vertebral bodies can present with a mixture of upper and lower neuron findings.
LMN, lower motor neuron; UMN, upper motor neuron.

INTERNATIONAL STANDARDS FOR NEUROLOGIC CLASSIFICATION OF SPINAL CORD INJURY

Neurologic Examination and Definitions

- Documenting impairments in a person with an SCI is best determined by performing a standardized neurologic examination as endorsed by the International Standards for Neurological Classification of Spinal Cord Injury (ISNCSCI).
- The examination is composed of sensory and motor components and is recommended to be performed with the patient in the supine position to be able to compare initial and follow-up exams.
- The information from this examination is recorded on a standardized flow sheet (**Figure 7–14**), and helps determine the sensory, motor, and neurologic level of injury (NLI); sensory and motor index scores; and to classify the impairment using the AIS.

1. 🕮 Sensory Testing

- For the sensory examination, there are 28 key sensory *dermatomes* that are individually tested for *light touch* and *pinprick* modalities. The face is used as the normal control point.
- Abnormal sensory score should be tagged with a "*" to indicate an impairment due to a non-SCI condition. The non-SCI condition should be explained in the comments box together with information about how the score is rated for classification purposes (normal/not normal for classification).
- Note: One of the key changes in the 2019 update to the International Standards is how to document exam findings that are not related to SCI and expanded the use of "*" to describe any sensorimotor impairments that are thought to be due to a non-SCI condition (e.g., pain, fracture, burn, peripheral nerve injury, etc.). In these situations, **the examiner should record the actual examined scores and tag them with an "*."** The non-SCI condition is then documented in the comments box with the recommendation for how the "*"-tagged scores should be treated during classification. This includes whether they are considered normal (typically rostral to the NLI) or abnormal (at or caudal to the NLI) for classification.
- Example of sensory "*": A person with light touch testing as 1 or altered at L4, L5, S1 dermatome, but due to diabetic neuropathy and not SCI, is designated as 1* for these levels, and in the comment box documented that this is due to a non-SCI issue and should be recorded as normal for classification.
- If accurate sensory testing is unable to be performed at the key sensory point due to extenuating circumstances (e.g., burns, casts, amputations, etc.), the level is designated as not testable, or "NT," on the worksheet, or an alternate location within the dermatome can be tested with a notation in the comments box that an alternate site was used.

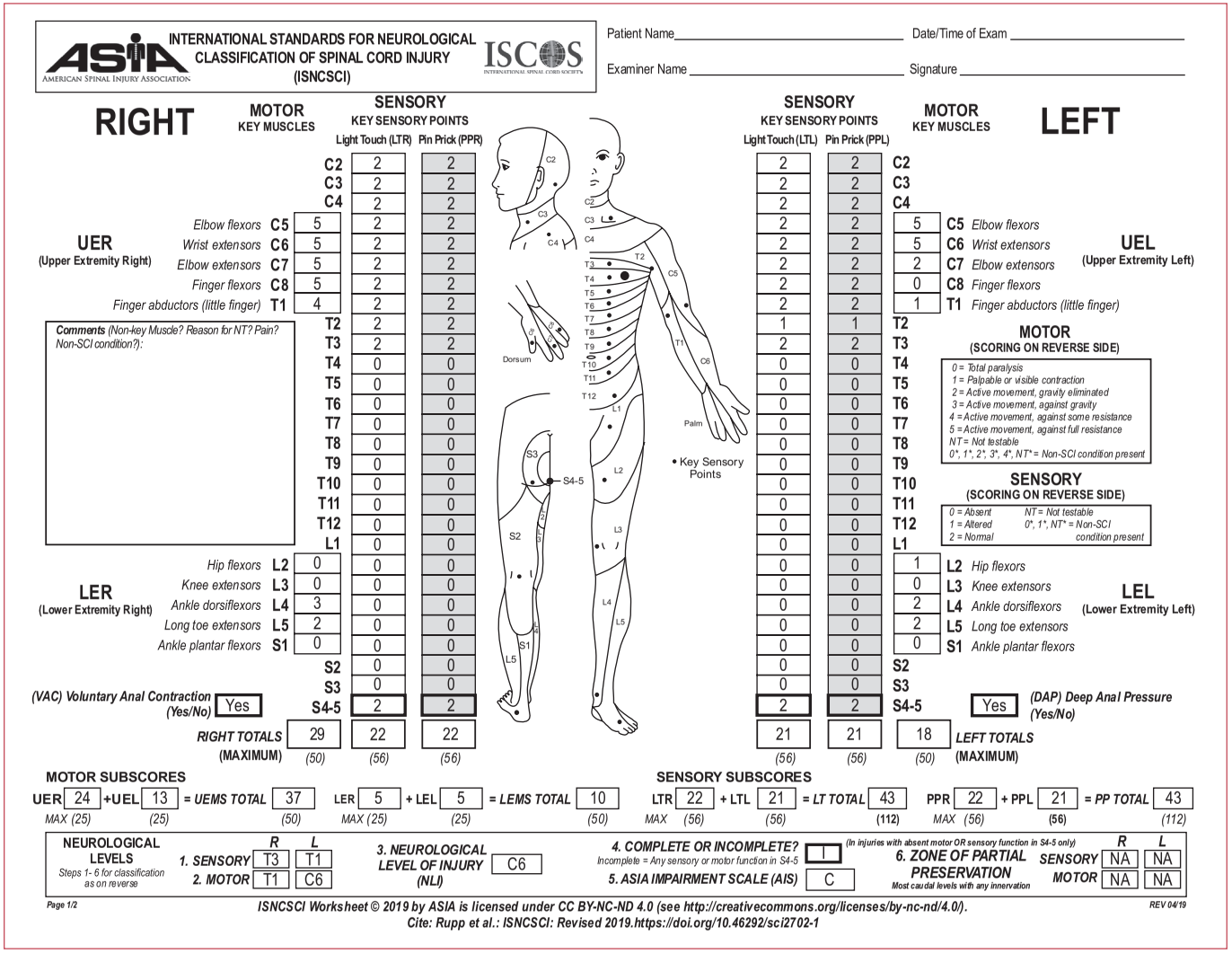


FIGURE 7–14 Scoring of a hypothetical patient with a C6 motor incomplete injury. *Source:* From American Spinal Injury Association. International Standards for Neurological Classification of Spinal Cord Injury. https://asia-spinalinjury.org/wp-content/uploads/2019/04/ASIA-ISCOSIntlWorksheet_2019.pdf. Updated April 2019. © 2019 American Spinal Injury Association. Reprinted with permission.

SCORE	SENSATION
0	Absent
1	Altered, either decreased/impaired sensation or hypersensitivity
2	Normal
NT	Not testable
0*, 1*, NT*	Non-SCI condition present

- If there is a question whether the patient can definitively discriminate between the sharp and dull edges, 8/10 correct answers is considered accurate, as this reduces the probability of correct guessing to <5%.
- **Pinprick testing:** Using the sharp edge of a *safety pin*, sensation is compared with that on the face. The patient must be able to differentiate the sharp and dull edge of a safety pin.

Score	0	No sensation at all or unable to differentiate between the sharp and dull edge
	1	The pin is not felt as sharply as on the face, but able to differentiate sharp from dull. (This difference can be hypo- or hyperesthesia.)
	2	Pin is felt as sharp as on the face.

- **Light touch testing:** A cotton tip applicator is used and sensation is compared with the face.

Score	0	Absent sensation
	1	Impaired—light touch is felt but different than on the face
	2	Normal—same as on the face

- 🕮 It is very important to test the S4–S5 dermatome for light touch and pinprick sensation, as this is a crucial part of what is used to distinguish between a neurologically complete and incomplete injury.

KEY SENSORY LEVELS

C2	1 cm lateral to occipital protuberance
C3	Supraclavicular fossa
C4	Superior AC joint
C5	Lateral side of the antecubital fossa
C6	Thumb–dorsal surface, proximal phalanx
C7	Middle finger–dorsal surface, proximal phalanx
C8	Little finger–dorsal surface, proximal phalanx
T1	Medial (ulnar) side of antecubital epicondyle
T2	Apex of axilla
T3	Third ICS at mid-clavicular line
T4	Nipple line—fourth ICS at mid-clavicular line
T5	Fifth ICS—midway between T4 and T6 at mid-clavicular line
T6	Xiphisternum—sixth ICS at mid-clavicular line
T7	One-quarter distance T6 and T10—at mid-clavicular line
T8	Midway between T6 and T10—at mid-clavicular line
T9	Three-quarter distance T6 and T10—at mid-clavicular line
T10	Umbilicus—at mid-clavicular line
T11	Midway between umbilicus and inguinal ligament—at mid-clavicular line
T12	Inguinal ligament at midpoint

(*continued*)

KEY SENSORY LEVELS (*continued*)

L1	Half the distance between T12 and L2
L2	Mid-anterior thigh
L3	Medial femoral condyle
L4	Medial malleolus
L5	Dorsum of foot at third MTP joint
S1	Lateral heel
S2	Popliteal fossa in the midline
S3	Ischial tuberosity
S4–S5	Perianal area (taken as one level)

AC, acromioclavicular; ICS, intercostal space; MTP, metatarsophalangeal.

- **Sensory level of injury:** Most caudal segment of the spinal cord with normal (2/2 score) sensory function on both sides of the body for both pinprick and light touch sensation
- If sensation is abnormal at C2, the sensory level is designated as C1.
- If sensation is intact through S4–S5, the sensory level should be recorded as intact ("INT") rather than as S4–S5.
- If the patient is unable to reliably appreciate sensation when tested on the face, then "NT" should be recorded, and "ND" (not determinable) should be documented in the appropriate area on the worksheet with no sensory level given.
- The presence of "*"-tagged sensory scores may influence the classification of levels, completeness, AIS grade, and zones of partial preservation (ZPP). If the classification of any of these variables is based on a clinical assumption, the classification component should be tagged with an "*."
- ***Sensory index scoring*** is calculated by adding the scores for all dermatomes, for a possible total score of 112 (56 on each side) for pinprick and light touch.

2. 🕮 Motor Strength Testing

- There are 10 key myotomes (referred to as the **key muscle groups**) tested on the left and right sides of the body in the supine position (see Table 7–2).

TABLE 7–2 Key Myotomes

MYOTOME	KEY MUSCLE FUNCTION
C5	Elbow flexors (Biceps brachialis)
C6	Wrist extensors (Extensor carpi radialis)
C7	Elbow extensors (Triceps)
C8	Finger flexors (FDP of middle finger)
T1	Small finger abductor (Abductor digiti minimi)
L2	Hip flexors (Iliopsoas)
L3	Knee extensors (Quadriceps)
L4	Ankle dorsiflexors (Tibialis anterior)
L5	Long toe extensors (Extensor hallucis longus)
S1	Ankle plantar flexors (Gastrocnemius)

FDP, flexor digitorum profundus.

- Example of "*": A person with manual muscle testing function having 3/5 strength at C5 motor level but due to brachial plexus injury and not SCI is designated as 3*, and in the comment box documented that this is due to a non-SCI issue and should be recorded as normal for classification.
- **Motor level of injury:** Most caudal key muscle function that is graded ≥3/5 with all the segments above graded 5/5 in strength. Motor level can be determined for each side of the body.
- For myotomes that are not clinically testable by manual muscle testing (i.e., above C5, T2–L1, and S2–S5), they are assumed to have full innervation if sensory innervation for pinprick and light touch at the corresponding level is also intact.
  - In the case where the C5 motor function is graded at least 3 on both sides of the body, with a sensory level on the right of C3 and on the left of C4, with impaired sensation at C4 on the right, the motor level on the right would be C3 (and C5 on the left).

MANUAL MUSCLE TESTING GRADING SYSTEM

0	No movement, total paralysis
1	Palpable or visible contraction
2	Active movement through full ROM with gravity eliminated
3	Active movement through full ROM against gravity
4	Active movement against moderate resistance through full ROM
5	Normal strength based on age, sex, and body habitus
NT	Not testable (e.g., due to immobilization, severe pain such that the patient cannot be graded, amputation of limb, or contracture of >50% ROM)
0*, 1*, 2*, 3*, 4*, NT*	Non-SCI condition present[a]

[a]Abnormal motor scores should be tagged with a "*" to indicate an impairment due to a non-SCI condition. The non-SCI condition should be **explained in the comments box** together with information about **how the score is rated for classification purposes** (normal/not normal for classification).
ROM, range of motion; SCI, spinal cord injury.

  - If, for example, all upper limb key muscle functions are normal, with intact sensation to T6, the motor level defers to the sensory level and is recorded as T6. However, with a T6 sensory level, if the T1 muscle function is graded a 3 (or a 4) instead of a 5, while T6 is still the sensory level, the motor level would be T1, as all the muscle levels above the T6 level are not normal.
- If "NT" has been documented as part of the exam, and this muscle is required for determination of the motor level, the designation of the motor level for that side should be deferred and ND (not determinable) is documented on the worksheet.
- The presence of "*"-tagged motor scores may influence the classification of the motor level, as well as subsequently the AIS grade and ZPP. If the classification of any of these variables is based on a clinical assumption, the classification component should be tagged with an "*."
- **Motor index scoring:** A possible total score of 100 can be obtained when adding the muscle scores of the key muscle groups (25 points per extremity). It is recommended that the motor score should be separated into two scores, one composed of the 10 upper limb muscle functions and one of the 10 lower limb muscle functions, with a maximum score of 50 each.

3. Anal Exam

- Two components are tested during the anal examination: Deep anal pressure (DAP) and voluntary anal contraction (VAC).
- DAP is tested by inserting a lubricated gloved finger into the anus with pressure applied to the anorectal wall using the thumb to gently squeeze the anus against an inserted index finger.
  - The patient is asked if they can appreciate this digital pressure.
  - Consistently perceived pressure is recorded as either present (YES) or absent (NO) on the worksheet.
- VAC is tested by inserting a lubricated gloved finger into the anus and asking the patient to "squeeze my finger as if to hold back a bowel movement."
  - This is graded as either present (YES) or absent (NO) in the appropriate box on the worksheet.
  - Care must be taken during this exam for patient modesty, as well as to differentiate volitional contraction from anal spasm when the finger is inserted or anal contraction is triggered by Valsalva.

4. 🕮 Neurologic Level of Injury

- Most caudal segment of the spinal cord with both normal sensory and motor function ≥3/5 with cephalad segments graded 5/5 on both sides of the body
- The motor and sensory levels are the same in <50% of complete injuries.

- In cases where there is no key muscle level available (i.e., cervical levels at and above C4, T2–L1, and sacral levels below S2), the NLI is that which corresponds to the sensory level, **if testable motor function above that level is also normal**.
  - Example: A person with the C5 muscle having 2/5 strength with a C4 sensory level would be assigned a motor level and NLI of C4. Additionally, a person with the same motor strength and a C3 sensory level would be assigned a motor level of C3.
  - If any sensory or motor level is determined based on "*" from examiner, then the neurologic level should also be marked with "*."

5. Skeletal Level of Injury

- Level where the greatest skeletal damage is noted by radiographic evaluation

NEUROLOGICALLY COMPLETE VERSUS INCOMPLETE SPINAL CORD INJURY

- SCI can be classified as neurologically "complete" or "incomplete" based upon sparing of function in the lowest sacral segments. Classification is dependent on the presence or absence of *sacral sparing* **(Table 7–3)**.
- 🕮 **Sacral sparing:**
  - Sacral sparing is evaluated by testing voluntary anal sphincter contraction (VAC) and sensory function (light touch and pinprick sensation at the S4–S5 dermatome, and DAP on rectal examination). Sparing of sacral sensation includes having impaired or intact sensation on either side, or DAP or VAC present. Even one of these being present is enough to be considered having sacral sparing.
  - The presence of sacral sparing represents at least partial structural continuity of the white matter long tracts. There is better prognosis for motor and sensory return below the level of injury as well as the possibility of return of bowel and bladder function in a person with sacral sparing relative to persons without sacral sparing (i.e., a neurologically complete injury).

🕮 **TABLE 7–3 ASIA Impairment Scale**

A = Complete	**No motor or sensory function (sensory and motor complete) is preserved in the sacral segments S4–S5.**
B = Sensory Incomplete	**Sensory is preserved but not motor function (motor complete) below the neurologic level and at the most caudal sacral segments S4–S5 (light touch or pinprick at S4–S5 or DAP), AND no motor function is preserved more than three levels below the motor level on either side of the body.**
C = Motor Incomplete	**Motor function is preserved at the most caudal sacral segments (S4–S5) on VAC OR the patient meets the criteria for sensory incomplete status (sensory function preserved at the most caudal sacral segments [S4–S5] by LT, *sharp/dull discrimination* or DAP), with sparing of motor function more than three levels below the ipsilateral motor level on either side of the body. This includes key or non-key muscle functions more than three levels below the motor level to determine motor incomplete status. For AIS C—less than half of key muscle functions below the single NLI have a muscle grade ≥3.**
D = Motor Incomplete	**Motor incomplete status as defined earlier, with at least half (half or more) of key muscle functions below the single NLI having a muscle grade ≥3.**
E = Normal	**If sensation and motor function as tested with the ISNCSCI are graded as normal in all segments, and the patient had prior deficits, then the AIS grade is E. Someone without an initial SCI does not receive an AIS grade.**

🕮 *Note:* For an individual to receive a grade of C or D, that is, motor incomplete status, they must have either (a) VAC or (b) sacral sensory sparing with sparing of motor function more than three levels below the motor level for that side of the body. The International Standards allow non-key muscle function more than three levels below the motor level to be used in determining motor incomplete status (AIS B vs. C).
🕮 *Note:* When assessing the extent of motor sparing below the level for distinguishing between AIS B and C, the *motor level* on each side is used. To differentiate between AIS C and D (based on proportion of key muscle functions with strength grade 3 or greater), the *single neurologic level* is used.
AIS, ASIA Impairment Scale; ASIA, American Spinal Injury Association; DAP, deep anal pressure; ISNCSCI, International Standards for Neurological Classification of Spinal Cord Injury; NLI, neurologic level of injury; SCI, spinal cord injury; VAC, voluntary anal contraction.
Source: From American Spinal Injury Association. International Standards for Neurological Classification of Spinal Cord Injury. Updated April 2019. https://asia-spinalinjury.org/wp-content/uploads/2016/02/International_Stds_Diagram_Worksheet.pdf, reprinted, with permission.

Complete Spinal Cord Injury

- **No sacral sparing:** Absence of sensory and motor function in the lowest sacral segment (S4–S5 dermatome)
  - "N-0-0-0-0-N" in the sacral exam noted on the worksheet is also known as noon sign.

"N-0-0-0-0-N" Mnemonic	
	N - No VAC
Then S4–S5	**0** (Light touch on the right)
	0 (Pinprick on the right)
	0 (Light touch on the left)
	0 (Pinprick on the left)
And S4–S5	**N** - No DAP

DAP, deep anal pressure; VAC, voluntary anal contraction.

Incomplete Spinal Cord Injury

- **Presence of sacral sparing** indicates at least partial preservation of sensory and/or motor function below the NLI that includes the lowest sacral segments. Sacral sensation and motor function are assessed.
- Again, sacral sparing (incomplete SCI) represents at least partial structural continuity of the white matter long tracts (i.e., corticospinal and spinothalamic tracts). There is a better prognosis for motor and sensory return below the level of injury as well as the possibility of return of bowel and bladder function relative to persons with a neurologically complete injury.
- 🕮 Classifying an AIS level: The AIS classifies the injury.

***STEPS IN CLASSIFYING THE INJURY ACCORDING TO THE AMERICAN SPINAL INJURY ASSOCIATION IMPAIRMENT SCALE* (ASIA ISCOS, 2019):**

1. **Determine *sensory levels* for right and left sides:**
   - The sensory level is the most caudal, intact dermatome for both pinprick and light touch sensation. Last level with "2" for light touch and pinprick.
2. **Determine *motor levels* for right and left sides:**
   - The motor level is the most caudal key muscle function that is graded ≥3/5 with all segments above graded 5/5 strength.
   - In regions where there is **no myotome** to test, the motor level is presumed to be the same as the sensory level, if testable motor function above that level is also normal.
3. **Determine the NLI:**
   - The most caudal segment of the cord with intact sensation and antigravity (3 or more) muscle function strength, provided that there is normal (intact) sensory and motor function rostrally
   - The most cephalad of the sensory and motor levels determined in steps 1 and 2
4. **Determine whether the injury is complete or incomplete (sacral sparing):**
   - If VAC = No AND all S4–S5 sensory scores = 0 AND DAP = No, then injury is **complete.**
   - Otherwise injury is **incomplete.**
5. **Determine AIS grade:**
   a. **Is injury complete**?
      - If Yes, AIS = A (no sacral sparing; no motor or sensory function left below level of injury)
      - If No, incomplete

   b. **If incomplete, is injury motor complete**?
      - If Yes, AIS = B (sensory function preserved below neurologic level + sacral sparing; no motor function)
      - If No, motor incomplete (no VAC or motor function more than three levels below the motor level on a given side)

c. **If motor incomplete,** are ≥50% of the key muscles below the **neurologic level** graded 3 or better?
   - If No, AIS = C
   - If Yes, AIS = D

d. If **sensation and motor function are normal in all segments, AIS = E.**
   - Note: AIS E is used in follow-up testing when an individual with a documented SCI has **recovered normal function**.
   - If **no deficits are found at initial testing**, the individual is considered to be neurologically intact, and the AIS **does not apply**.

6. Determine the ZPP.
   – The term ZPP is only used in injuries with absent motor (no VAC) OR sensory function (no DAP, no LT, and no PP sensation) in the lowest sacral segments S4–S5, and refers to those dermatomes and myotomes caudal to the sensory and motor levels that remain partially innervated. With sacral DAP sparing of sensory function, or in cases with no PP or LT on that side, the sensory ZPP is not applicable and "NA" is used. In a case of no DAP present, for instance, and PP or LT absent on one side, the one side without any sparing will have a ZPP, but the other side would record "NA." For motor, if VAC is present, the motor ZPP is not applicable ("NA").

STEPS FOR CLASSIFICATION OF INJURY IN CASES WITH A ""-TAGGED SCORE*

1. Perform the classification replacing the "*"-tagged scores with the assumed scores.
2. Record the classification results based on the assumed scores (which include the examiner's judgment) in the respective classification boxes at the bottom of the worksheet.
3. Next, reclassify the case with the examined scores (while ignoring the "*"-tags).
4. Tag each classification variable which is different between classification with the assumed scores and reclassification with the examined scores with an "*" to indicate that it is based on clinical assumptions.

- **Non-key muscles** refer to muscle functions that are distinct from the key muscle functions that are routinely tested and can help distinguish between AIS B and C (see Table 7–4). In a patient with an apparent AIS B classification, with no key muscle function present, non-key muscle functions >3 levels below the motor level on each side should be tested to most accurately classify the injury and differentiate between an AIS B versus C.
- Table 7–4 lists the non-key muscles and their innervations/levels that should be used for them.
- For pediatric patients, training for the examination is described in the WeeSTeP (www.asia-spinalinjury.org/learning/). The comprehensive examination of the ISNCSCI is thought to be too complex for the cognitive abilities and tolerance of children younger than 6 years old, and some patients as old as 8 may have difficulty with the exam.

CLINICAL PRESENTATION OF SPINAL SHOCK

Stage of Spinal Shock

- 🕮 **Spinal shock** is a temporary loss or depression of all spinal reflex activity below the level of the lesion, although this may not occur in all patients.
- There is a loss of motor function and sensation accompanied by atonic paralysis of the bladder and bowel.
- Muscles below the level of the lesion become flaccid and hyporeflexic.
- Autonomic function below the level of the lesion is also impaired. Temporary loss of piloerection, sweating, and vasomotor tone in the lower parts of the body.

Reflexes Returning After Spinal Shock

- **Delayed plantar response:**
  – Usually the first to return after spinal shock (Ko, 2018)
  – Elicited by stroking the sole of the foot in the lateral plantar aspect beginning at the heel, moving up to the ball of the foot, staying lateral to the great toe. Same area as for the Babinski sign but requires deep pressure rather than a light stimulus.

TABLE 7–4 Non-Key Muscle Groups

MOVEMENT	ROOT LEVEL
Shoulder: Flexion, extension, abduction, adduction, internal and external rotation **Elbow:** Supination	C5
Elbow: Pronation **Wrist:** Flexion	C6
Finger: Flexion at proximal joint, extension **Thumb:** Flexion, extension, and abduction in plane of thumb	C7
Finger: Flexion at MCP joint **Thumb:** Opposition, adduction, and abduction perpendicular to palm	C8
Finger: Abduction of little finger	T1
Hip: Adduction	L2
Hip: External rotation	L3
Hip: Extension, abduction, internal rotation **Knee:** Flexion **Ankle:** Inversion and eversion **Toe:** MTP and IP extension	L4
Hallux and toe: DIP and PIP flexion and abduction	L5
Hallux: Adduction	S1

DIP, distal interphalangeal; IP, interphalangeal; MCP, metacarpophalangeal; MTP, metatarsophalangeal; PIP, proximal interphalangeal.

  - The response is delayed in comparison to a normal plantar response (first movement of great toe is flexion, adduction of the other toes) or Babinski sign.
  - The toes flex and then relax slowly.
  - If persistent, there is a high correlation with complete injuries with poor prognosis for lower extremity (LE) recovery.
- **Bulbocavernosus reflex (BCR; Figure 7–15):**
  - After the delayed plantar response, the BCR returns soon after injury (usually within 24 hours).
  - The absence of the reflex in a person with acute paralysis from trauma indicates the presence of spinal shock or a possible LMN injury.
  - Performed by squeezing the tip of the penis (in men) or the clitoris (in women) or tugging on a Foley catheter and noting stimulation of anal sphincter contraction
- **Perianal sphincter reflex (anal wink):**
  - Perianal stimulation causes contraction of the anal sphincter.
  - Indicates similar findings to the BCR

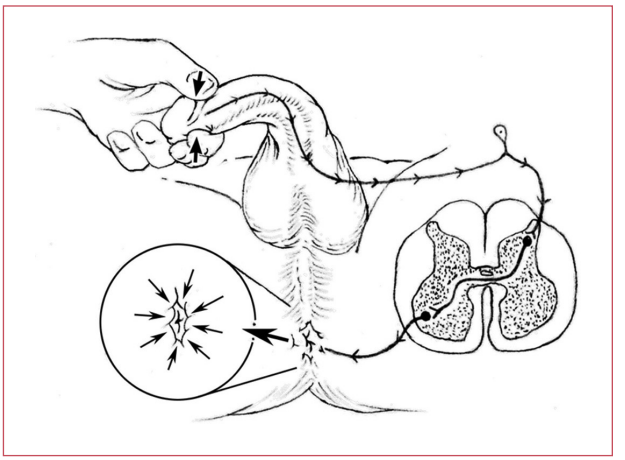


FIGURE 7–15 The bulbocavernosus reflex.

🕮 Duration/Phases of Spinal Shock (Ditunno et al., 2004)

- **Phase 1 (0–24 hours)**: Areflexia of all reflexes below SCI level, which typically lasts for 24 hours. Reflexes begin to return within 24 hours.
- **Phase 2 (days 1–3)**: Initial reflex activity is noted, usually with the return of the delayed plantar response followed by the BCR and the anal wink. A positive Babinski sign may be noted at a later time.
- **Phase 3 (day 4–1 month)**: Early hyperreflexia—on average, muscle stretch reflexes return after 2 to 3 weeks. Some reflexes (e.g., bladder) may not return for up to 3 months (or later) after injury.
- **Phase 4 (1–12 months)**: Spasticity/hyperreflexia—muscle stretch reflexes become hyperactive with the presence of pathologic reflexes below the lesion, resulting in spasticity.
- **Spinal shock can last days to weeks to months**.
- The loss of deep tendon reflexes (spinal shock) on average lasts 4 to 12 weeks (Ko, 2018).

INCOMPLETE SPINAL CORD INJURY SYNDROMES

🕮 Central Cord Syndrome

- Most common of the incomplete SCI syndromes
- Produces greater motor weakness in the upper limbs than the lower limbs, with variable loss of sensation, bowel, and bladder function
- Historically, it was felt that CCS affected the central aspects of the spinal cord, with the neuroanatomy of the CST having a cervical distribution medially and sacral distribution laterally, thereby affecting the UEs more than LEs. The understanding of the pathophysiology of CCS has changed since the initial description of CCS. Studies have demonstrated a CCS is often related to a diffuse white matter injury to the lateral columns and CST, rather than the central cord. It is also now apparent that the CST is not somatotopically organized, and is critical for hand function but less so for ambulation.
- May occur at any age but is more common in older patients with cervical spondylosis who sustain a cervical hyperextension injury, usually from a fall
- 🕮 Recovery: LEs recover first and to a greater extent. This is followed by improvement in bladder function, then proximal UE, and finally intrinsic hand function.
- Age below 50 and the degree of motor sparing initially are key positive prognostic indicators of functional recovery.

🕮 Brown-Séquard Syndrome (Figures 7–16 and 7–17)

- Results from a lesion that causes a relative hemisection of the spinal cord
- Rare injury that constitutes 2% to 4% of all traumatic SCI
- Associated classically with stabbing but can occur from other causes (e.g., MVC)
- Neurologic deficits distal to the level of the lesion vary from the different nerve tracts crossing at different locations:

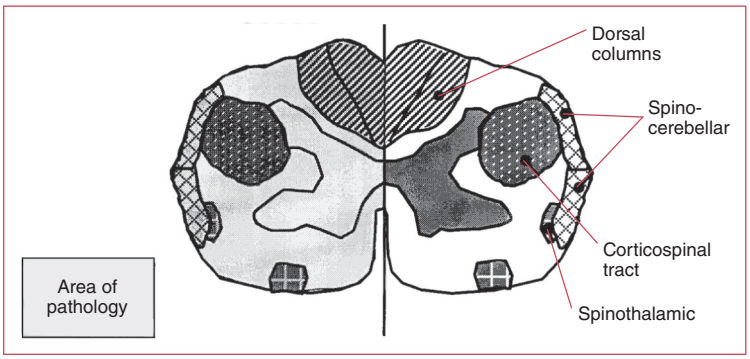


FIGURE 7–16 Brown-Séquard syndrome. Transverse section of the spinal cord—refer to Figure 7–2 for anatomic landmarks.

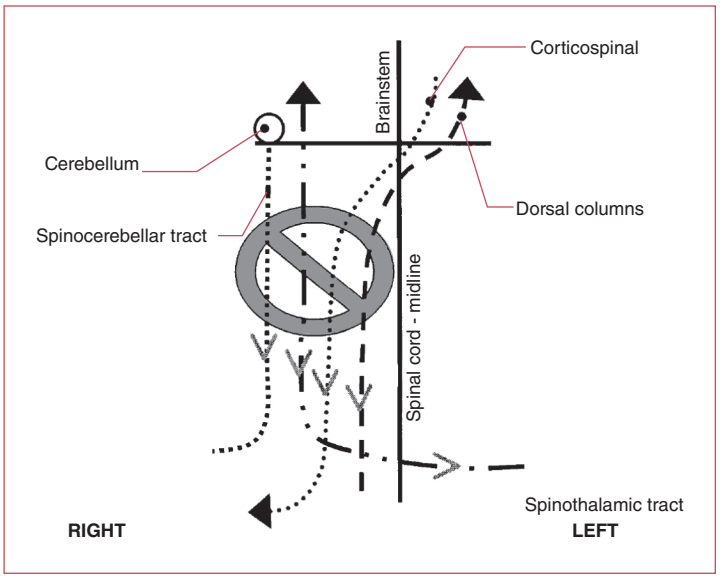


FIGURE 7–17 Brown-Séquard syndrome lesion: Depicts point of injury, that is, right-sided knife wound. Follow tracts distal from the point of injury. Result is ipsilateral motor and proprioceptive deficits (right-sided), and contralateral pain and temperature deficits (left-sided).

  - At the level of lesion:
    - Ipsilateral flaccid paralysis (anterior horn cells)
    - Ipsilateral loss of all sensory modalities
  - Below the level of lesion:
    - Ipsilateral paralysis (CST)
    - Ipsilateral loss of light touch and proprioception (dorsal columns)
    - Contralateral loss of pain and temperature (spinothalamic tract)
- Overall, patients clinically present most often with a relative ipsilateral motor and proprioceptive loss, and contralateral loss of pain and temperature.
- Usually a very good prognosis for recovery of ambulation

🕮 Anterior Cord Syndrome (Figure 7–18)

- A lesion affecting the anterior two-thirds of the spinal cord while preserving the posterior columns
- This can occur from flexion injuries, direct injury to the anterior spinal cord from bone fragments or disc herniation, or anterior spinal artery lesions.

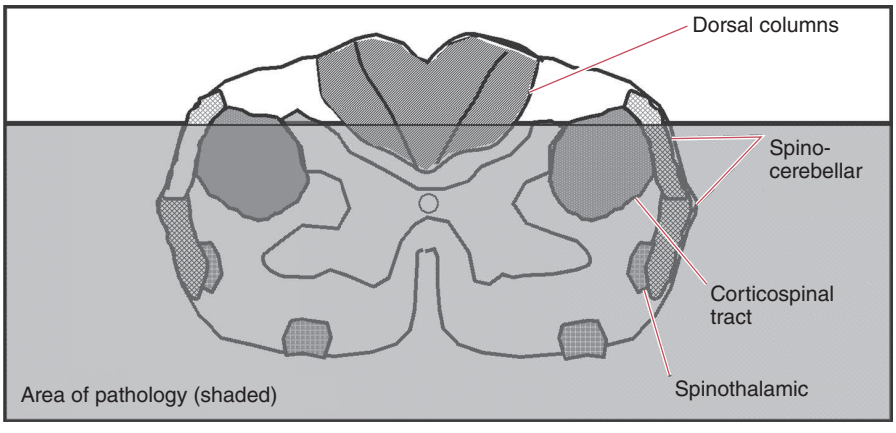


FIGURE 7–18 Anterior cord syndrome. Transverse section of the spinal cord—refer to Figure 7–2 for anatomic landmarks.

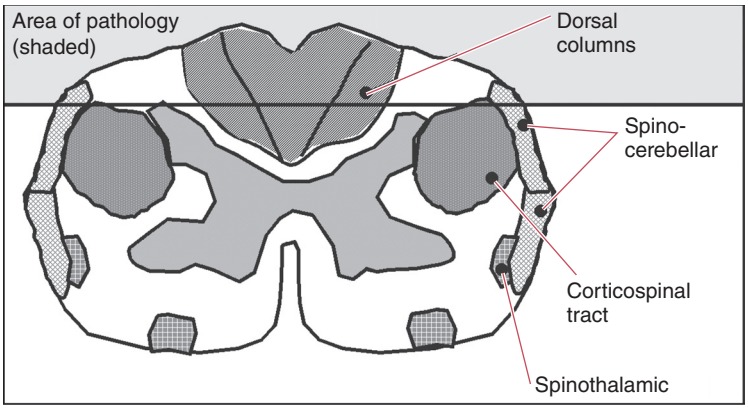


FIGURE 7–19 Posterior cord syndrome. Transverse section of the spinal cord—refer to **Figure 7–2** for anatomic landmarks.

- Results in:
  - Variable loss of motor function (CST)
  - Variable loss of pain, temperature, and pinprick sensation (spinothalamic tract)
  - Preservation of proprioception, light touch, and deep pressure sensation (dorsal columns)
  - 🕮 Only 10% to 20% chance of muscle recovery
  - Even in individuals with some recovery, motor strength is typically low and coordination is lacking, causing low chance of walking recovery.
- Prognostic factors:
  - Etiology of injury is primary factor. Aortic dissection/rupture and high cervical lesions have greater risk of death.
  - More severe initial presenting symptoms at initial presentation have poorer outcomes.
  - Lack of improvement in the initial 24 hours indicates poorer outcome.
  - Female sex and old age correlate with poorer outcomes.

Posterior Cord Syndrome (Figure 7–19)

- This has been omitted from recent versions of the International Standards because of its rare presentation after traumatic injury (least common syndrome, <1% incidence).
- Injury to the posterior columns results in proprioceptive loss (dorsal columns) with muscle strength, pain, and temperature modalities spared.
- Prognosis for ambulation is poor secondary to the proprioceptive deficits.

Conus Medullaris Syndrome (Figure 7–20 and Table 7–5)

- The conus medullaris, which is the terminal segment of the adult spinal cord, typically lies at the inferior aspect of the L1–L2 vertebrae.
- If it is a high conus lesion such as that in **Figure 7–20A**, BCR and micturition reflexes are preserved.

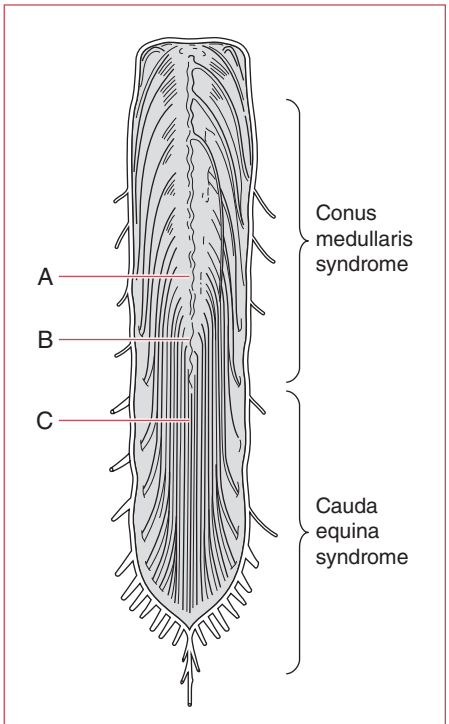


FIGURE 7–20 Distal spinal cord: (A) High conus medullaris lesion. (B) Low conus medullaris lesion. (C) Cauda equina lesion affecting the lumbosacral nerve roots.
Source: From American Spinal Injury Association©, 1996, with permission.

TABLE 7–5 Conus Medullaris Versus Cauda Equina Syndrome

CONUS MEDULLARIS SYNDROME L1–L2 VERTEBRAL LEVEL	CAUDA EQUINA SYNDROME BELOW L2 TO SACRAL VERTEBRAL LEVEL
Location: T12–L1–L2 vertebral level injury of sacral cord and lumbosacral nerve roots	**Location:** Below L1–L2 to sacrum vertebral level injury of lumbosacral nerve roots
Causes: • T12–L1 fracture • Trauma • Infarction • Tumors • Epidural abscess • Spinal hemorrhage • Arteriovenous malformation • Spina bifida, tethering of the cord • Disc herniation	**Causes:** • Fracture at L1–L2 or below with significant retropulsion, including sacral fracture • Large disc herniation • Severe spinal stenosis • Trauma • Spinal hemorrhage • AVM • Tumor • Infection
Resultant Signs and Symptoms: 1. Back pain with radiating LE pain 2. Symmetric LE weakness 3. Sensory loss in LEs and saddle anesthesia 4. A high conus lesion typically presents as a UMN injury (hyperreflexia, spasticity) 5. A lower conus lesion may present as a UMN, LMN, or mixed picture (hyper- and hyporeflexia) 6. Bowel, bladder, and sexual dysfunction may occur	**Resultant Signs and Symptoms:** 1. Back pain with or without radiating LE pain 2. Leg weakness 3. Sensory loss/numbness in LEs (based on root distribution) and saddle anesthesia 4. Results in an LMN lesion – Flaccid paralysis of LEs – Hyporeflexia or areflexia in affected muscle groups 5. Abnormalities may be asymmetric 6. High cauda equina lesions (lumbar roots): – Spare bowel and bladder 7. Low cauda equina lesions (S3–S5): – Causes areflexic bowel, bladder, and sexual dysfunction
EMG: Normal EMG (except for EAS or S1, S2 involvement).	**EMG:** Findings show multiple spinal nerve root involvement (polyradiculopathy)

AVM, arteriovenous malformation; EAS, external anal sphincter; EMG, electromyography; LE, lower extremities; LMN, lower motor neuron; UMN, upper motor neuron.

- Most commonly, injury occurs at the distal part of the conus as depicted in **Figure 7–20B**. At this level, the conus and lumbar nerve roots within the spinal canal are injured, resulting in areflexic bladder and bowel, and lower limb weakness (in low-level lesions). This results in a mixed presentation of UMN and LMN findings.

🕮 Cauda Equina Syndrome (Figure 7–20 and Table 7–5)

- Injuries below the L1–L2 vertebral levels affecting the cauda equina (nerve rootlets) as seen in **Figure 7–20C**, which innervate the lumbar and sacral segments. The cauda equina is composed of spinal nerve rootlets, which are peripheral nerves, and would result in LMN signs and symptoms.
- Results in motor weakness and atrophy of the LEs (L2–S2) with neurogenic bowel and bladder (S2–S4), sexual dysfunction, and areflexia below the level of injury (including absence of anal wink/BCR).
- Overall, has a better prognosis relative to UMN injuries for recovery, most likely due to the fact that the nerve roots (peripheral nerves) are more resilient to injury and more likely to regenerate (Bulloch et al., 2022)

FUNCTIONAL OUTCOMES AFTER SPINAL CORD INJURY

- The most important factors in determining functional outcome are the motor level and the degree of impairment, that is, AIS classification (Tables 7–6 and 7–7).

TABLE 7–6 Potential Functional Outcomes for Persons With Complete Tetraplegia

ACTIVITY	C1–C4	C5	C6	C7	C8–T1
Feeding	Dependent	Mod-Ind after setup	Mod-Ind	Mod-Ind	Independent
Grooming	Dependent	Min assist with equipment after setup	CG–Mod-Ind	Mod-Ind	Independent
UE dressing	Dependent	Mod-Max assist	Mod-Ind	Independent	Independent
LE dressing	Dependent	Dependent	Min-Mod assist	Mod-Ind–CG	Mod-Ind
Bathing	Dependent	Mod-Max assist	Min assist with equipment	CG–Mod-Ind with equipment	Mod-Ind
Bed mobility	Dependent	Mod–Max assist	CG–CS	Mod-Ind	Independent
Weight shifts	Mod-Ind in power WC; Dependent in manual WC	Mod-Ind in power WC; Mod assist in manual WC	Independent	Independent	Independent
Transfers	Dependent	Mod–Max assist	CG on level surfaces	Independent with or without board for level surfaces	Independent
WC propulsion	Mod-Ind with power WC; Dependent in manual WC	Mod-Ind in power WC; Mod-Ind in power assist WC; Mod-Ind in manual WC on level surfaces	Independent in manual WC with coated rims on level surfaces	Independent except curbs and uneven terrain	Independent
Driving	Modified passenger van	Mod-Ind with van conversion	Mod-Ind with adaptations	Car with hand controls or adapted van	Car with hand control or adapted van
Bowel and bladder	Dependent	Dependent	Mod-Ind	Independent	Independent

CG, contact guard; CS, close supervision; LE, lower extremity; Max assist, maximum assistance; Min assist, minimum assistance; Mod assist, moderate assistance; Mod-Ind, modified independent; UE, upper extremity; WC, wheelchair.

TABLE 7–7 Potential Functional Outcomes for Persons With Complete Paraplegia

ACTIVITY	T2–T9	T10–L2	L3–S5
ADLs (grooming, feeding, dressing, bathing)	Independent	Independent	Independent
Bowel/bladder	Independent	Independent	Independent
Transfers	Independent	Independent	Independent
Ambulation	Exercise only	Household ambulation; can trial ambulation outdoors	Community ambulation is possible
Braces	Bilateral KAFOs; walker or forearm crutches	Bilateral KAFOs, forearm crutches	Possibly KAFO or AFOs; forearm crutches or canes

ADLs, activities of daily living; AFOs, ankle–foot orthoses; KAFO, knee–ankle–foot orthosis.

- Other factors such as age, body habitus, cognitive impairment, participation, and secondary complications such as pain and spasticity may also impact outcomes.
- Sensation and pinprick preservation may also be important predictors for motor recovery and ambulation post-SCI.
- 🕮 Some evidence shows that individuals with sacral pinprick preservation post-SCI along with LE PP sparing have higher walking outcomes for limited ambulation at 1 year.
- Functional goals should be individualized to meet each person's strengths, weaknesses, and individual circumstances.
- 🕮 The highest complete SCI level that can live independently without the aid of an attendant is a C6 complete tetraplegia:
  - This patient would have to be *extremely motivated*.
  - Feeding is accomplished with a universal cuff for utensils.
  - Transfers require stabilization of elbow extension with forces transmitted from shoulder musculature through a closed kinetic chain.
  - Bowel care is performed using a suppository inserter or other apparatus for digital stimulation.
- 🕮 However, **C7 level is the *usual* level for achieving independence**.
- Outcome studies of a subset of patients with motor and sensory complete C6 SCI revealed that for the majority of self-care tasks, including feeding, grooming, dressing, and transfers, <20% are independent.

MEDICAL COMPLICATIONS OF SPINAL CORD INJURY

🕮 **Important Levels to Remember**

- **T6 and above:** Individuals with SCI are considered to be at risk for BP dysregulation. This includes:
  - AD
  - Orthostatic hypotension
- **T8 and above:** If lesion above T8, patient cannot regulate and maintain normal body temperature.
  - Note: An easy way to remember this level is to spell the word "***temp-eight-ture.***"
  - Central temperature regulation in the brain is located in the hypothalamus.

AD, autonomic dysreflexia; BP, blood pressure; SCI, spinal cord injury.

🕮 ORTHOSTATIC HYPOTENSION (TABLE 7–8)

- State of transient reflex depression caused by a lack of sympathetic outflow and triggered by tilting the patient upright to >60 degrees
  - Lesion at T6 or above
  - T1–L2 responsible for tachycardia, vasoconstriction, and increased arterial pressure
  - Heart and blood vessels supplied by T1–T7
- **Mechanism:**
  - Upright position causes decrease in blood pressure (BP).
  - Aortic and carotid baroreceptors sense decrease in BP (would usually increase sympathetic outflow in neurologically intact individual); however, efferent pathway is interrupted following SCI.
  - Brainstem unable to send message through spinal cord to cause sympathetic outflow and vasoconstriction of splanchnic bed to increase BP

  - Orthostasis often lessens with time due to development of spinal postural reflexes, which allow for vasoconstriction, due to improved autoregulation of cerebrovascular circulation in the presence of perfusion pressure.
- **Symptoms:**
  - Lightheaded
  - Dizziness
  - Presyncope
  - Nausea
  - Pallor
- **Signs:**
  - Hypotension: Loss of sympathetic tone
    - Decreased systemic venous resistance, dilation of venous vessels → decreased preload to the heart
  - Tachycardia: Aortic and carotid baroreceptors respond to hypotension. However, interruption of efferent pathway precludes increase in sympathetic outflow.
    - Symptoms persist, however, because the increase in heart rate (HR) is not sufficient to counteract the decrease in BP.
  - Syncope
- 🕮 **Management:**
  - Reposition—Trendelenburg/recliner wheelchair (WC)
  - Elastic stocking/abdominal binder
  - Accommodation (e.g., use of tilt table)
  - Increase fluid intake
  - Pharmacologic agents:
    - Salt tablets 1 g four times a day
    - Midodrine (ProAmatine®; alpha-1 adrenergic agonist): 2.5 to 10 mg three times a day (TID)
    - Fludrocortisone (Florinef®; mineralocorticoid): 0.1 to 0.4 mg daily
    - Droxidopa (Northera®): Starting dose is 100 mg three times during the day, maximum dose 600 mg TID.
  - Caution: Once orthostasis improves, the patient may be at risk for AD.

🕮 AUTONOMIC DYSREFLEXIA (TABLE 7–8)

- **Mechanism:** Syndrome of massive imbalanced reflex sympathetic discharge in patients with SCI above the splanchnic outflow (T5–L2)
- Occurs in patients with lesions at T6 and above and affects 48% to 90% of susceptible patients. AD is secondary to the loss of descending central sympathetic control and hypersensitivity of receptors below the level of the lesion.
- **Pathophysiology:**
  - Noxious stimulus—increases sympathetic reflex spinal release
  - Regional vasoconstriction—causes a marked rise in arterial BP
  - Increases peripheral vascular resistance—increases cardiac output, increases BP
  - Aortic and carotid baroreceptors respond to increased BP and relay impulses to vasomotor center in brainstem—impulses via vagus nerve that can lead to bradycardia. However, this is not effective in combating the increased BP. Bradycardia, while classic, is not always seen, and tachycardia, including arrhythmias (i.e., atrial fibrillation), may occur more frequently.
  - 🕮 Note: The brainstem is unable to send messages through the injured spinal cord to decrease sympathetic outflow and allow vasodilation of splanchnic bed to decrease BP.
  - **Onset:** Occurs after spinal shock and may appear within 2 to 4 weeks postinjury. If it occurs in a patient, it will present within the first year in >90% of cases. More commonly occurs in patients with neurologically complete SCI (apt to have more severe symptoms), although it may occur in patients with incomplete SCI.
- **Cause:** Noxious stimulus below the level of the lesion
  - Most commonly from bladder (overdistention or infection), followed by bowel (fecal impaction)

TABLE 7–8 Orthostatic Hypotension Versus Autonomic Dysreflexia

ORTHOSTATIC HYPOTENSION	AUTONOMIC DYSREFLEXIA
Trigger: Tilting patient >60 degrees toward the upright	**Trigger:** Noxious stimulus below level of lesion, especially full bladder or bowel
Due to: Lack of sympathetic outflow **Lesion:** T6 or above	**Due to:** Too much sympathetic outflow, loss of descending control, hypersensitivity **Onset:** Status post spinal shock usually within first 6 months **Lesion:** T6 or above
Symptoms: Lightheadedness/dizziness, syncope **Signs:** Hypotension due to upright positioning **Tachycardia:** Aortic and carotid baroreceptors respond to hypotension	**Symptoms:** Headache Sweating above level of SCI Flushing above level of SCI Piloerection Pupillary constriction Sinus congestion **Signs:** • HTN (systolic BP >20 mmHg above baseline) • Bradycardia: Aortic and carotid baroreceptors respond to HTN • Tachycardia may present more frequently
Etiology: Upright position causes decrease in BP. Aortic and carotid baroreceptors sense the decrease in BP; however, brainstem is unable to send message through SC to cause sympathetic outflow and subsequent vasoconstriction of splanchnic bed to increase BP.	**Etiology:** Noxious stimulus causes massive sympathetic output. Aortic and carotid baroreceptors sense increased BP; however, brainstem is unable to send message through SC to decrease sympathetic outflow and allow for vasodilation of splanchnic bed to bring BP down.
Treatment: 1. Reposition back toward Trendelenburg 2. Elastic stockings 3. Abdominal binders 4. Fluids 5. Medications: – Salt tablets – Midodrine (alpha-1 adrenergic agonist) – Fludrocortisone (Florinef®; mineralocorticoid) – Droxidopa (Northera®)	**Treatment:** 1. Sit patient up 2. Remove noxious stimulus (look for bladder distension, fecal impaction, etc.) 3. Treat HTN: – Consider temporary treatment with nitropaste (transdermal), clonidine (oral), nifedipine (oral), or hydralazine. – Occasionally may require IV agents, such as diazoxide, nitroprusside, hydralazine, and spinal anesthesia – It is estimated that 48% to 85% of patients with high-level SCI have symptoms of AD. – When untreated, can lead to: (1) Retinal hemorrhage (2) CVA (3) SAH, seizure, MI, death

AD, autonomic dysreflexia; BP, blood pressure; CVA, cerebral vascular accident; HTN, hypertension; IV, intravenous; MI, myocardial infarction; SAH, subarachnoid hemorrhage; SC, spinal cord; SCI, spinal cord injuries.

- **Most common causes:**
  - Bladder: Blocked catheter/distended bladder
  - Bowel: Fecal impaction
  - Abdominal emergency (appendicitis, cholecystitis, pancreatitis)
  - Labor
  - Pressure injuries

  - Fractures
  - Ingrown toenails
  - Orgasm
  - Urinary tract infections (UTIs)
  - Epididymitis
  - Bladder stones
  - Gastric ulcers
- **Signs and symptoms:**
  - Elevated BP (systolic BP >20 mmHg above baseline)
  - Headache
  - Sweating above level of SCI
  - Flushing above level of SCI
  - Piloerection
  - Pupillary constriction
  - Sinus congestion
- 🕮 **Management:**
  1. Sit patient upright (the first aspect of treatment) and loosen all tight-fitting clothing and devices (e.g., elastic band from urine leg bag, elastic stockings, abdominal binder).
  2. 🕮 Identify and remove noxious stimulus—early bladder evaluation (flush indwelling catheter if present; catheterize patient if needed; use intravesical lidocaine if present prior to urinary tract manipulation to lessen further noxious stimulation).
  3. Monitor BP every 2 to 5 minutes during the episode and monitor for recurrent symptoms for at least 2 hours after resolution to ensure that it does not recur.
  4. Medications should be initiated if BP is significantly elevated (>150 mmHg) and one is unable to find source quickly; should be started prior to checking for fecal impaction in the event that early bladder survey yields no improvement in BP/symptoms.
- **Pharmacotherapy:**
  - *Acute (several options used):*
    - Nitropaste: ½ in. (to start) up to 2 in., place above the level of injury and should be removed once noxious stimulus is corrected
    - Oral clonidine: 0.1 to 0.3 mg
    - Nifedipine (Procardia®): 10 mg chew and swallow
    - Hydralazine
  - *ICU management:* A number of medications can be used to decrease the BP, including:
    - Diazoxide
    - Nitroprusside
    - Labetalol
- **Prevention:** Only occasionally required. Options include alpha- and beta-blockers.
  - 🕮 Some evidence shows prazosin given prior to common triggers may prevent episode of AD.
- 🕮 **Pregnancy/surgery:** Spinal anesthesia is recommended during delivery with SCI at T6 or above.
- **Potential complications of AD:**
  - If hypertensive episodes are not treated, complications can potentially lead to:
    - Retinal hemorrhage
    - Cerebral vascular accident (CVA)/subarachnoid hemorrhage (SAH)
    - Seizure
    - Myocardial infarction (MI)
    - Death
  - AD predisposes the patient to cardiac dysrhythmias (e.g., atrial fibrillation) by altering the normal pattern of repolarization of the atria, making the heart susceptible to reentrant-type arrhythmias.

BLADDER DYSFUNCTION

Neuroanatomy and Neurophysiology of Voiding

CENTRAL PATHWAYS

- **Frontal lobe (Corticopontine mesencephalic nuclei):**
  - Inhibits parasympathetic sacral micturition center
  - Allows bladder storage
- **Pons (Pontine mesencephalic nuclei):**
  - Coordinates bladder contraction and sphincter relaxation
  - Loss of control from this center can result in detrusor sphincter dyssynergia (DSD).
- **Pelvic and pudendal nuclei: Sacral micturition:**
  - Integrate stimuli from cephalic centers
  - Mediate parasympathetic sacral (S2–S4) micturition reflex
- **Motor cortex to pudendal nucleus:**
  - Voluntary control (contraction/inhibition) of external urethral sphincter

PERIPHERAL PATHWAYS (FIGURE 7–21)

- Control of micturition in the peripheral nervous system is coordinated by interactions between the autonomic nervous system (sympathetic and parasympathetic) and somatic nervous system.
- Micturition in the autonomic nervous system is a balance between the sympathetic and parasympathetic nervous systems:
  - Sympathetic nervous system will result in urine storage (think "fight or flight" response).
  - Parasympathetic nervous system will cause urine release (relaxation response) and originates in the sacral micturition center and innervates the detrusor muscle.
- Somatic efferent nerve fibers from motor cortex to pudendal nucleus in the sacral micturition center give voluntary control (contraction/inhibition) of the external urethral sphincter.
- **Parasympathetic efferents:**
  - *Origin:* Detrusor nucleus in intermediolateral gray matter at S2–S4 levels
  - *Course:* Travel through **pelvic nerves** to parasympathetic receptors of detrusor muscle
  - *Function:* Stimulation of cholinergic receptors causes bladder contraction and emptying.
- **Sympathetic efferents:**
  - *Origin:* Intermediolateral gray matter from T11–L2
  - *Course:* Travel through **hypogastric nerves** to alpha-1 and beta-2 adrenergic receptors within the bladder and urethra
  - *Function:* Stimulation of beta-3 adrenergic receptors within the body of the bladder causes smooth muscle relaxation (compliance) + stimulation of alpha-1 adrenergic receptors within the base of the bladder/prostatic urethra causes smooth muscle contraction (increase outlet resistance). This combination of actions *results in* **urine storage**.
- **Somatic efferents:**
  - *Origin:* Pudendal nucleus of sacral segments (S2–S4)
  - *Course:* Travel through pudendal nerve to innervate striated muscle of external urethral sphincter
  - *Function:* Voluntary contraction of external urethral sphincter **prevents leakage or** emptying.
- **Afferent fibers:**
  - *Origin:* Detrusor muscle stretch receptors, external anal and urethral sphincters, perineum, genitalia
  - *Course:* Travel through the pelvic and pudendal nerves to the sacral cord
  - *Function:* Myelinated A-delta fibers respond to bladder distention stimulating parasympathetic emptying of bladder. Unmyelinated C-fibers are silent and not essential for normal voiding; however, increased activity can be seen following SCI (target of capsaicin and resiniferatoxin to control neurogenic detrusor overactivity [NDO]).

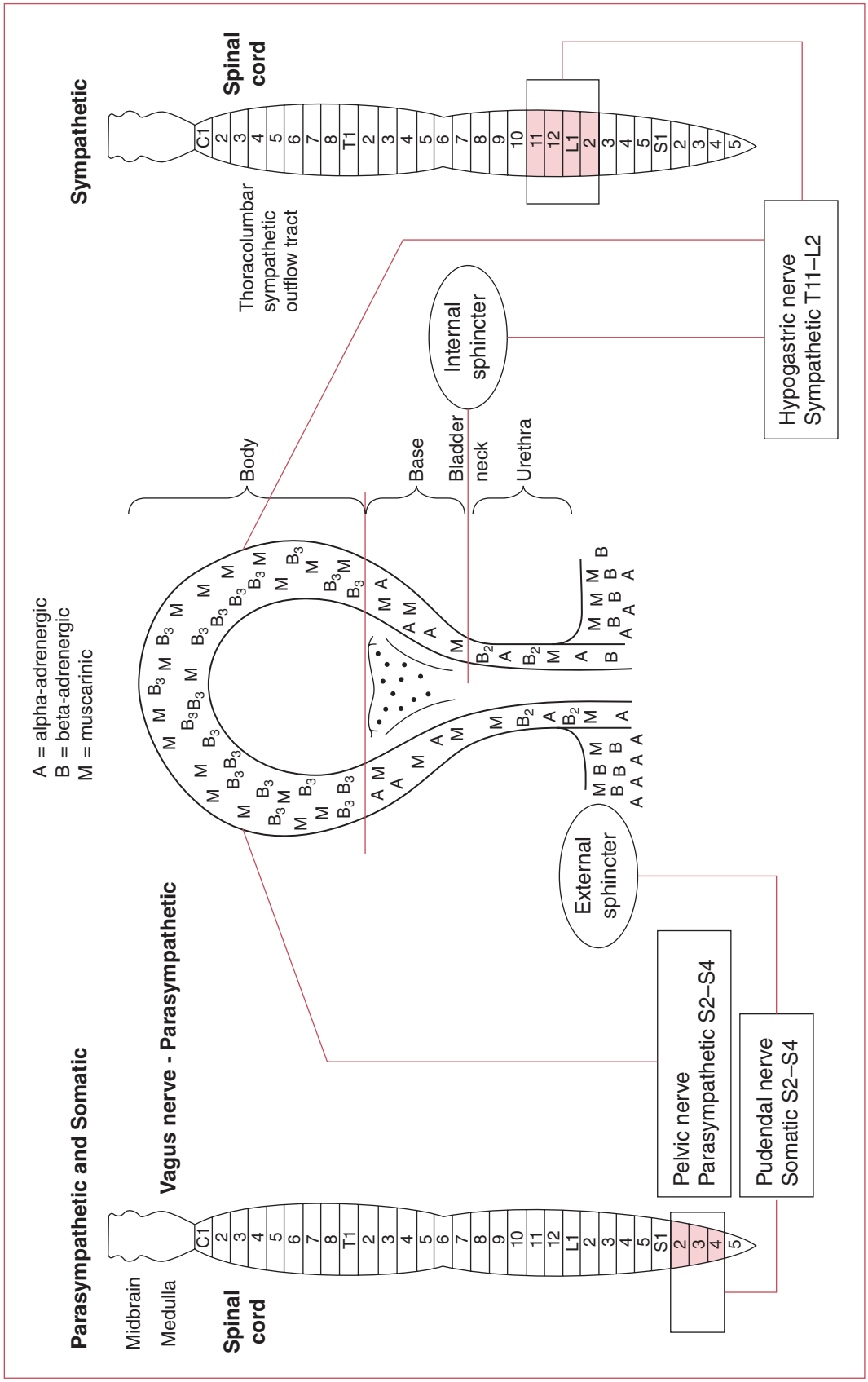


FIGURE 7–21 Neurologic innervation and receptors of the bladder and sphincter.

URETHRAL SPHINCTERS

- **Internal sphincter:**
  - Mostly innervated by T11–L2 hypogastric nerve (sympathetic)
  - Under control of autonomic system; large number of alpha-adrenergic receptors
  - Contracts sphincter for storage
  - Smooth muscle, involuntary
- **External sphincter:**
  - Innervated by pudendal nerve (S2–S4)
  - Prevents leakage or emptying
  - Skeletal muscle, voluntary

Bladder Receptors (see Figure 7–21)

- **Cholinergic muscarinic receptors (M3):**
  - Located within the bladder wall, trigone, bladder neck, and urethra
  - Acetylcholine (Ach) binds to M3 receptors to cause contraction
- **Beta-2 adrenergic receptors:**
  - Activate relaxation of the bladder neck to promote emptying
- **Beta-3 adrenergic receptors:**
  - Concentrated in the body of the bladder, also some in bladder neck
  - Norepinephrine (NE) binds to beta-adrenergic receptors to cause relaxation
- **Alpha-1 adrenergic receptors:**
  - Located within the base of the bladder and prostatic urethra
  - NE binds to alpha-1 adrenergic receptors to cause contraction
- 🕮 Note: The bladder wall does not have baroreceptors.

- **Alpha**-adrenergic receptors respond to the appearance of NE with *contraction.*
- **Beta**-adrenergic receptors respond to the appearance of NE with *relaxation.*

NE, norepinephrine.

NORMAL BLADDER FUNCTION: STORAGE VERSUS EMPTYING

Normal Bladder Storage

SYMPATHETIC RESPONSE (ENCOURAGED DURING "FIGHT OR FLIGHT" RESPONSE)

- Sympathetic tone predominates to promote internal sphincter contraction and bladder relaxation allowing for urine storage.
- *T11–L2 sympathetic efferents:*
  - Travel through the hypogastric nerves to activate alpha-1 and beta-3 adrenergic receptors
  - Causes sphincter to contract and body to relax
  - Urine is stored
- 🕮 *Activation of alpha-1 adrenergic receptors:*
  - Causes contraction of the internal sphincter at the base of the bladder and prostatic urethra, preventing leakage
  - Promotes storage
- 🕮 *Activation of beta-3 adrenergic receptors* (Yamaguchi & Chapple, 2007):
  - Causes relaxation of body of bladder to allow expansion
  - Promotes storage

Normal Bladder Emptying

PARASYMPATHETIC RESPONSE (ENCOURAGED DURING RELAXATION)

- Parasympathetic tone predominates causing bladder contraction and emptying.
- *S2–S4 parasympathetic efferents:*
  - Travel through the pelvic nerves to activate cholinergic (muscarinic M2) receptors
  - Ach stimulates cholinergic receptors in the bladder wall, trigone, neck, and urethra, causing bladder contraction and emptying

- *Beta-2 adrenergic receptors:*
  - Activated by NE upon initiation of voiding to cause relaxation of bladder neck
  - Promotes emptying

Note: Remember, **S**ympathetics "**S**tore" and **P**arasympathetics "**P**ee."

EVALUATION OF URINARY FUNCTION

History

- Should include present bladder management, fluid intake/output, and urologic symptoms (urgency, frequency, hesitancy, dysuria, incontinence, AD)
- Functional history is also very important (hand function, dressing skills, sitting balance, transfer ability, mobility status).

Physical Exam

- Should focus on determination of the NLI, abdomen, external genitalia, and sacral/perineal skin
- Anal sphincter tone and the presence/absence of sacral reflexes should be noted:
  - BCR (S2–S4)
  - Anal reflex (wink; S2–S4)

Laboratory Evaluation

- Serum creatinine level is not sensitive in detecting early deterioration of renal function in persons with SCI.
- 24-hour creatinine clearance is a more accurate laboratory method in this population.

Imaging

- Evaluation of function:
  - Upper tract: Quantitative renal MAG3 scan
- Evaluation of anatomy:
  - Upper tract:
    - Renal ultrasound (US)
    - CT*
    - MRI*
  - Lower tract:
    - Bladder US
    - Cystogram
    - Cystoscopy (not an imaging technique but will give information regarding bladder anatomy)

*Primarily used to evaluate anatomy but can give some information regarding renal function and drainage

Urodynamic Evaluation

- Evaluates two phases of bladder function: Filling (storage) and voiding (emptying)
- A typical urodynamic study for a person with normal bladder function is depicted in **Figure 7–22**.
- Filling:
  - Evaluates sensation, bladder capacity, bladder wall compliance, and bladder stability (the presence/absence of NDO)
  - Sensations evaluated include:
    - First sensation 100 to 200 mL
    - Sensation of fullness 300 to 400 mL
    - Sensation of urgency 400 to 500 mL
- Normal bladder capacity ranges between 400 and 500 mL

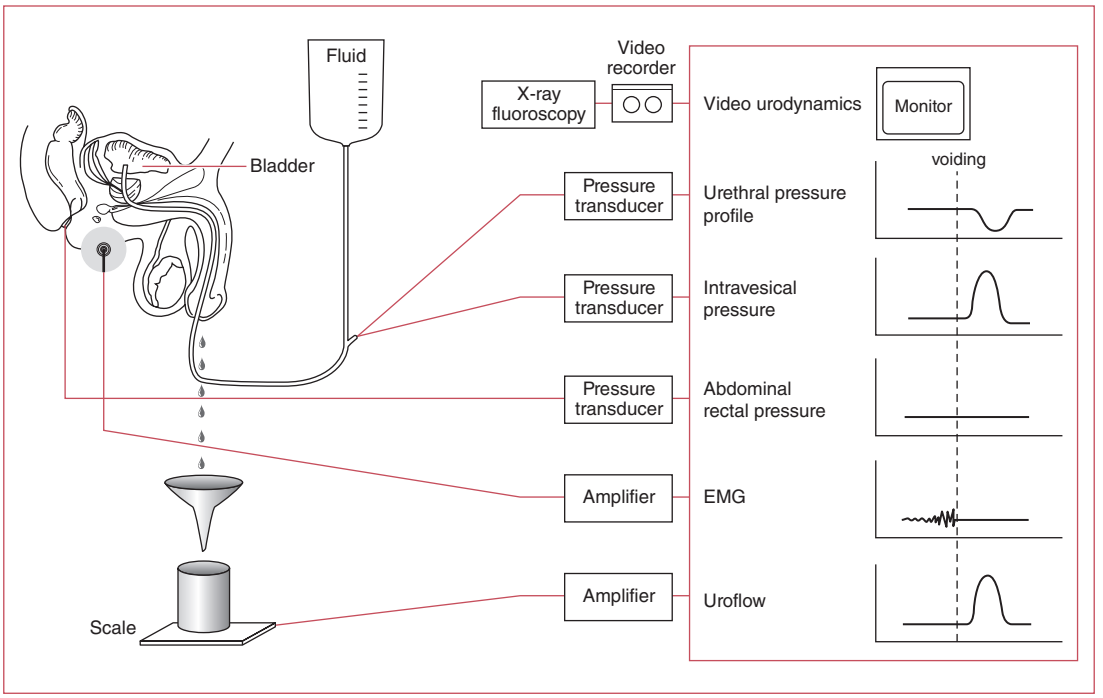


FIGURE 7–22 Instrumentation for urodynamic studies is not standardized. The illustration uses radio-opaque fluid. Normal bladder function can be divided into storage and voiding phases. The first sensation of bladder filling is between 100 and 200 mL. The able-bodied individual experiences bladder fullness between 300 and 400 mL and the sense of urgency between 400 and 500 mL. In a urodynamic study of a person with normal bladder function, intravesical pressure does not increase significantly during the storage phase due to the viscoelasticity of the vesical wall. During the voiding phase, sphincter activity stops, and the bladder contracts. During normal voiding, the EMG signal will be silent, intravesical pressure will increase, and urethral pressure will decrease. There should also be no rise in intra-abdominal pressure at any time during normal voiding. Fluoroscopy can be used for evaluation of VUR and does give an anatomic visualization of simultaneous bladder and sphincter function.
EMG, electromyography; VUR, vesicoureteral reflux.
Source: Adapted from Nesathurai S, ed. *The Rehabilitation of People with Spinal Cord Injury: A House Officer's Guide*. 3rd ed. Arbuckle Publishers; 2013.

- Emptying:
  - Evaluates detrusor, sphincter, and abdominal pressures/activity; urine flow; and postvoid residual
- *Functional bladder capacity* = voided volume + residual urine volume

Normal Detrusor Contraction Detrusor Pressures and Pelvic Floor Electromyography (Figure 7–23)

🕮 POST-SPINAL CORD INJURY UROLOGIC FUNCTION AND MANAGEMENT

- Previously termed as neurogenic bladder, the current terminology is neurogenic lower urinary tract dysfunction (NLUTD) as there is dysfunction of detrusor, sphincter, and pelvic floor.
- Goals:
  - Prevention of upper tract complications (e.g., deterioration of renal function, hydronephrosis, renal calculi, pyelonephritis)
  - Prevention of lower tract complications (e.g., cystitis, bladder stones, vesicoureteral reflux [VUR])
- Initial management:
  - Acutely, patients in spinal shock typically present with an *areflexic* bladder, which retains urine. This can last from a week to many months, but most cases resolve in 2 to 12 weeks.

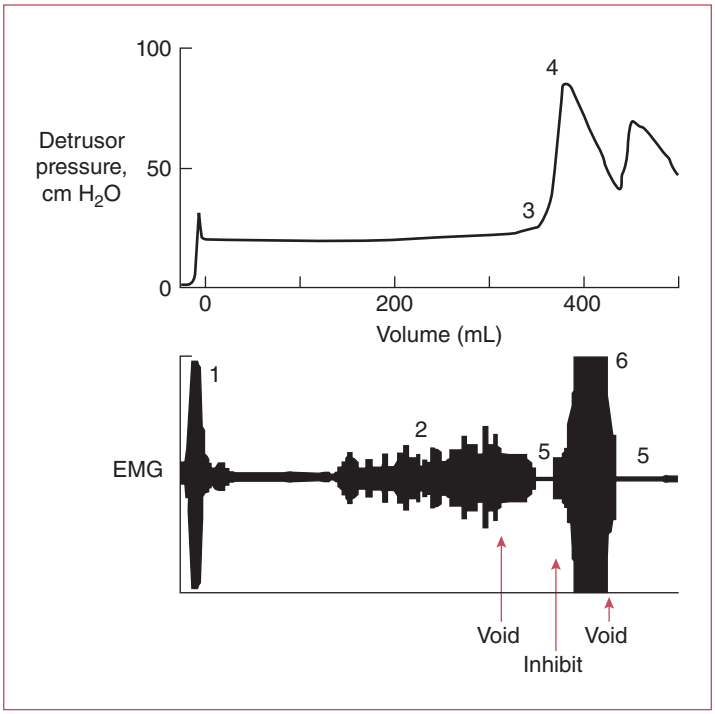


FIGURE 7–23 Normal cystometrogram/pelvic floor EMG. 1. Bulbocavernosus reflex. 2. Contraction of pelvic floor muscles during later phase of filling (progressively increasing electrical activity). 3. Functional bladder capacity. 4. Detrusor contraction that occurs during voiding. 5. Electrical silence (abrupt) that occurs during voiding. 6. Electrical activity of pelvic floor muscles that occurs during voluntary inhibition.
EMG, electromyography.

  - Indwelling catheter, especially while IV fluids are administered. Allows for urine removal and monitoring of urine output.
  - An intermittent catheterization (IC) program can be established in appropriate patients once they are no longer receiving IV fluids, urine output has stabilized (<100 mL/hr), and an approximate 2 to 3 L/day fluid restriction can be maintained:
    - Best started every 4 hours with goal catheterization volumes <500 mL
- Long-term management:
  - Based on type of bladder dysfunction and patient-specific factors (e.g., NLI, function, comorbidities, support)
  - Bladder dysfunction is often closely related to the level of injury (i.e., UMN vs. LMN injury); however, many variations exist.
  - Regular urodynamic studies should be performed to assess actual bladder function.
- Bladder dysfunction after SCI can be based upon the location of an injury:
  - Suprapontine lesions—a lesion from cerebral or brainstem with preservation of the pontine micturition center (e.g., stroke, traumatic brain injury [TBI])
  - Suprasacral spinal cord/pontine lesions—for example, SCI, TM
  - Sacral spinal cord lesions—for example, tumors, lumbar stenosis
  - Infraconal (cauda equina and peripheral nerve) lesions. An example can include diabetic neuropathy.
- However, many physicians still break the types of NLUTD into reflexive versus areflexive types of bladder. This is somewhat simplistic, as the suprapontine and suprasacral types of lesions both cause a reflexive bladder, yet the residual issues and treatments are not completely the same (Linsenmeyer, 2019).
- Bladder management programs can be categorized as follows:
  - Catheterization (e.g., Foley catheter, suprapubic catheter, or intermittent cath)
  - Bladder reflex triggering (reflex voiding)
  - Bladder expression (e.g., Valsalva's maneuver, Crede's maneuver), although these are not commonly recommended
  - Electrostimulation

TABLE 7–9 Neurogenic Lower Urinary Tract Dysfunction (NLUTD): Reflexic Upper Versus Areflexic Lower Motor Neuron

AREFLEXIC BLADDER/LMN BLADDER (FIGURE 7–24): FAILURE TO EMPTY	REFLEXIC BLADDER/UMN BLADDER (FIGURE 7–25): FAILURE TO STORE
Causes: • Commonly results from flaccid bladder and/or spastic sphincter • Lack of sacral reflexes • Spinal shock: Reflex arc not functioning due to initial trauma • Conus medullaris syndrome • Cauda equina syndrome • Syringomyelia • Acute CVA (detrusor areflexia)	**Causes:** • Commonly results from spastic bladder (detrusor hyperreflexia) and/or incompetent sphincter • NDO due to lack of inhibition of sacral micturition center • SCI: Return of reflex arc after initial trauma • Subacute CVA (detrusor hyperreflexia) • MS (most commonly see detrusor hyperreflexia)
Lesion: • Infrasacral lesion (i.e., involving sacral micturition center [S2–S4]) • Lesion exclusively involving the peripheral innervation of the bladder (LMN injury)	**Lesion:** • Lesion between the pons and sacral micturition center (above S2)
Can result in: • Large, areflexic, flaccid bladder • Failure to empty **Treatment:** • Catheterization (most common IC) • Crede maneuver (suprapubic pressure) not often recommended • Valsalva maneuver (need to make sure bladder pressures are safe when performing this technique) • Medications to promote emptying and induce urination: – Bethanechol: Stimulate cholinergic receptors in LMN injury; only helpful if some bladder contractions are present; rarely used in SCI	**Can result in:** • Small, overactive, spastic bladder • Failure to store or empty **Treatment:** • Drugs to allow for urine storage: – Anticholinergic medications (e.g., oxybutynin [Ditropan®], tolterodine [Detrol®], and others) – Stimulants of alpha-, beta-receptors to allow storage of urine are used (e.g., B3 agonists mirabegron [Myrbetriq®]) – Botulinum toxin to the detrusor

Note: It is important to remember that these are the *most commonly* seen types of bladder pathology in each case. However, several causes can fall into either category or display features of both LMN and UMN pathology at different times throughout the natural course of the disease (e.g., MS: Can see detrusor hyperreflexia, detrusor areflexia, or DSD; SCI or CVA: Can initially see detrusor areflexia followed by detrusor hyperreflexia).
CVA, cerebral vascular accident; DSD, detrusor sphincter dyssynergia; IC, intermittent catheterization; LMN, lower motor neuron; MS, multiple sclerosis; NDO, neurogenic detrusor overactivity; SCI, spinal cord injuries; UMN, upper motor neuron.

AREFLEXIC/LOWER MOTOR NEURON BLADDER (FIGURE 7–24): FAILURE TO EMPTY

- Internal sphincter may have increased tension and prevent voiding.
- When bladder can no longer expand, urine leakage occurs via *overflow incontinence*.

REFLEXIC/UPPER MOTOR NEURON BLADDER (FIGURE 7–25): FAILURE TO STORE

- Due to UMN lesion, there is *no* suppression of the sacral micturition center, resulting in an overactive, spastic bladder (detrusor hyperreflexia).
- As a result, the patient voids prematurely.

DETRUSOR SPHINCTER DYSSYNERGIA—COMBINATION TYPE BLADDER (FIGURE 7–26)

- Up to 85% of SCI patients develop DSD.
- Neurologic injury between the sacral (S2–S4) and pontine micturition centers (suprasacral spinal cord/pontine lesions) causes a lack of coordinated regulation of bladder function.

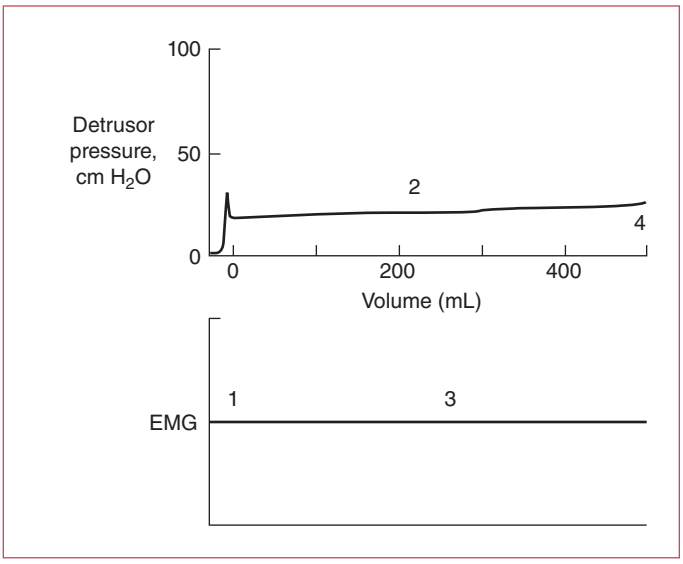


FIGURE 7–24 Cystometrogram and EMG in a patient with complete LMN bladder dysfunction. 1. Absent bulbocavernosus reflex. 2. Major detrusor contractions are absent. 3. No pelvic floor muscle activity (external urethral sphincter). 4. Large bladder capacity.
EMG, electromyography; LMN, lower motor neuron.

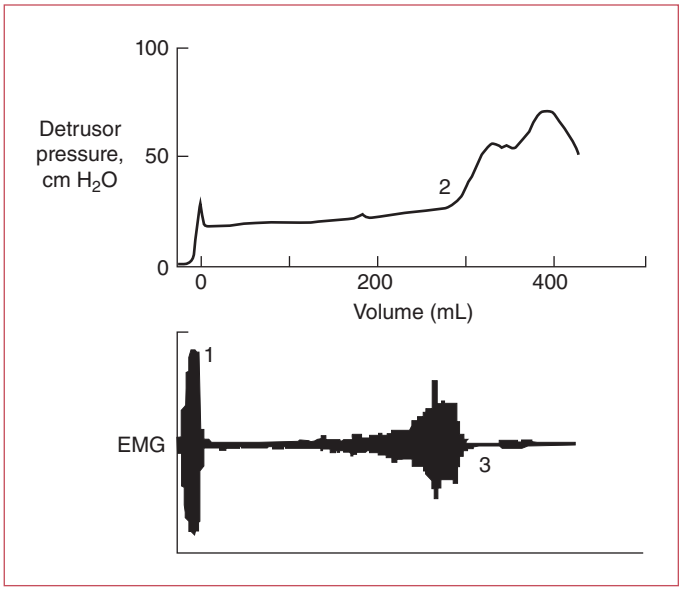


FIGURE 7–25 Cystometrogram and EMG in a patient with complete uninhibited neurogenic bladder lower urinary tract dysfunction. 1. Brisk bulbocavernosus reflex. 2. Spontaneous detrusor contraction at reduced bladder capacity. 3. Silence of pelvic floor muscles (external urethral sphincter).
EMG, electromyography.

- This can lead to above normal bladder pressures determined by urodynamic studies and should be treated to minimize renal dysfunction.

RESULT

- Small, overactive, spastic bladder (detrusor hyperreflexia)
- Tight, spastic external sphincter (sphincter hyperactivity)
- Overall result: Failure to empty and, if able to, will do so under high voiding pressures

RISK IF NOT TREATED

- VUR: Retrograde urine flow from the bladder toward the kidneys
- Result: Increased residual volumes, colonized infected stagnant urine, high-pressure voiding against a closed sphincter with risk of VUR

TREATMENT

1. Anticholinergic meds: Relax detrusor and suppress uninhibited bladder contractions, thereby preventing long-term complication of VUR (oxybutynin, tolterodine, trospium chloride, solifenacin, darifenacin, fesoterodine)

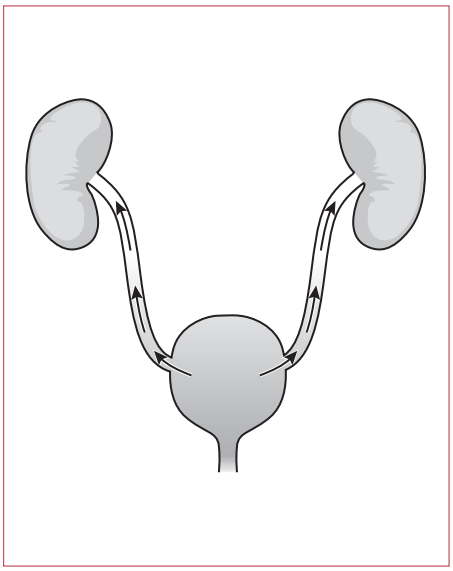


FIGURE 7–26A Reflux of infected urine backs up toward the kidneys.

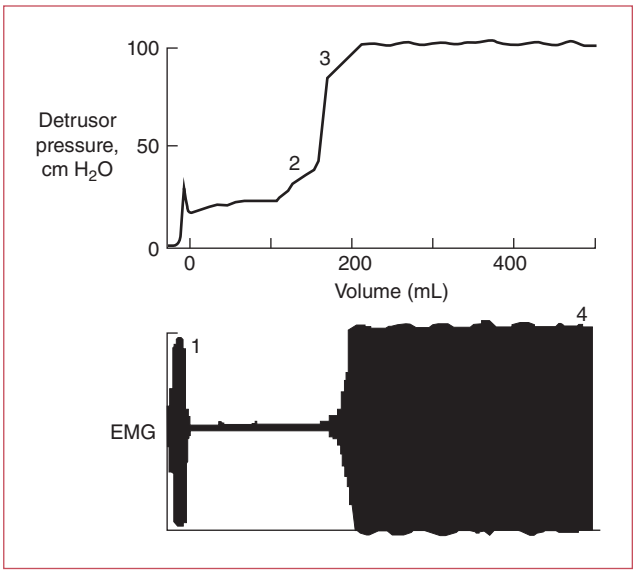


FIGURE 7–26B Cystometrogram and EMG in a patient with complete UMN bladder dysfunction: 1. Brisk bulbocavernosus reflex. 2. Bladder capacity is reduced. 3. High intravesical pressure during detrusor contraction. 4. Detrusor/external urethral sphincter dyssynergia with marked electrical activity of the pelvic floor muscles during detrusor contraction.
EMG, electromyography; UMN, upper motor neuron.

2. Beta-3 agonist: Sympathetic beta-3 adrenergic receptor agonist causes detrusor muscle relaxation (mirabegron).
3. IC
4. Botulinum toxin to detrusor wall
5. Alpha-blockers: Open bladder neck (alfuzosin, doxazosin, terazosin, tamsulosin, silodosin)
6. Sphincterotomy if considering reflex voiding

Pathophysiology of Vesicoureteral Reflux

NORMAL ANATOMY (FIGURE 7–27A1 AND A2)

- The one-way valve mechanism remains competent as long as the oblique course of the ureter within the bladder wall is maintained.
- During relaxation of the bladder, when urine is being stored, the ureter pumps urine into the bladder.
- During bladder contraction, the valve shuts closed. As a result, the bladder is emptied with no reflux of urine into the ureters.

ABNORMAL BLADDER ANATOMY (FIGURE 7–27B1 AND B2)

- Bladder wall hypertrophy causes the course of the distal ureter to become progressively perpendicular to the inner surface of the bladder. The vesicoureteral junction consequently becomes incompetent, permitting reflux of urine.
- During relaxation of the bladder, the ureter pumps urine into the bladder.
- The valve cannot close during bladder contraction given the perpendicular orientation of the distal ureter to the inner surface of the bladder. As such, urine is forced up the ureter to the kidney and hydronephrosis can result.
- Reflux can be further complicated by acute or chronic pyelonephritis with progressive renal failure.

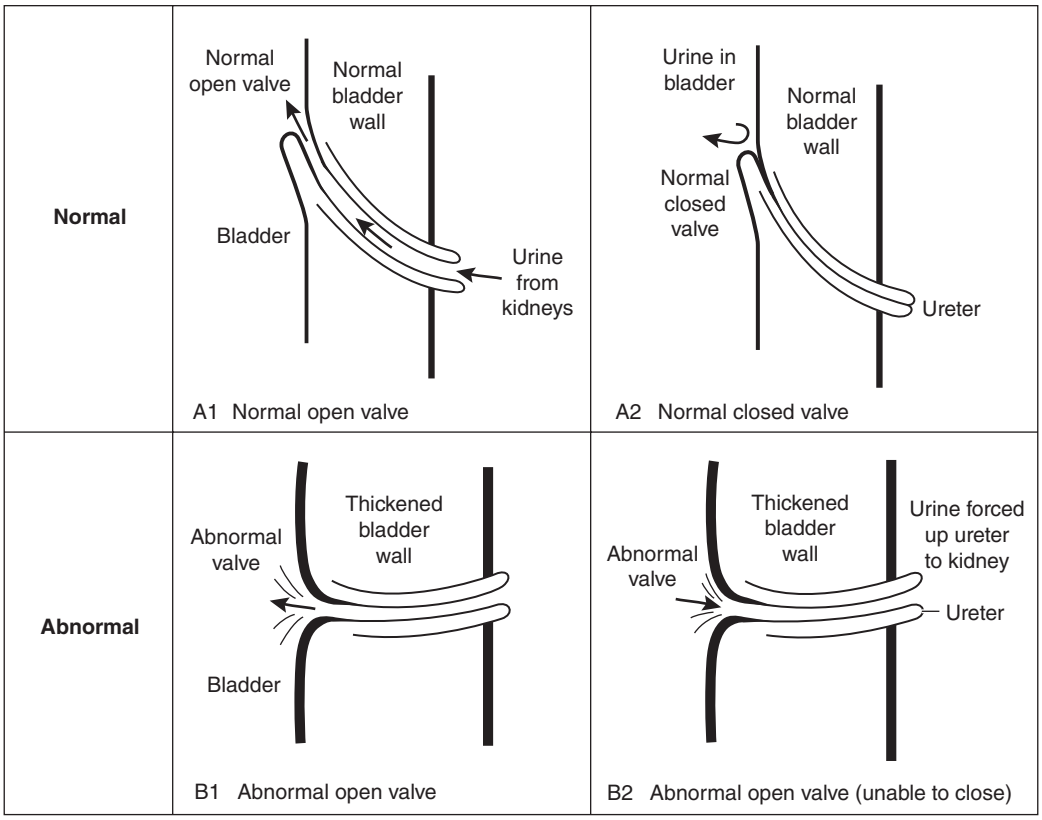


FIGURE 7–27 Vesicoureteral junction. (A): 1. Normal open valve: The muscle of the ureter milks urine through the valve into the bladder. 2. Normal closed valve: When the bladder contracts, the valve is pressed shut. The normal valve prevents urine from flowing back into the ureter. (B): 1. Abnormal open valve: The abnormal valve still allows urine to pass through the ureter into the bladder. 2. Abnormal valve: The abnormal valve is unable to close. When the bladder contracts, urine is pushed back into the ureter and kidney.

- Congenital abnormalities of the ureters, including posterior placed ureteral orifices and severe trabeculation disrupting the posterior bladder wall, have been associated with VUR after SCI.

Urinary Tract Complications Due to Neurogenic Lower Urinary Tract Dysfunction

- UTIs
- Bladder calculi
- VUR
- Hydronephrosis
- Renal deterioration
- Potentiation of AD

Urinary Tract Infections in Spinal Cord Injury

Pathophysiology

- UTIs are generally caused by the endogenous flora of the host overcoming the competing normal flora and the host defense mechanism.
- Acidic urine inhibits microbial growth. UTIs are prevented by the washout effort of large volumes of urine. The large flow of fluid impedes the adherence of microorganisms and dilutes their concentration.

- The presence of the UTI is affected by:
  - Virulence of the invading microorganism
  - Condition of the urine as the culture medium
  - Host defense mechanisms
- UTIs are the most common complication following SCI.
- Classified as asymptomatic or symptomatic UTI
- Asymptomatic UTI—not a true infection but instead colonization of the bladder
- Symptomatic UTI criteria:
  - Pyuria—increased white blood cells (WBCs) in the urine
  - Bacteriuria—increased bacterial counts in the urine
  - New onset of symptoms (e.g., cloudy urine, foul-smelling urine, dysuria, malaise, increased spasticity, worsening AD)

Management of Urinary Tract Infections

- **Asymptomatic UTIs**
  - In asymptomatic NLUTD patients, clinicians should not perform surveillance/screening urine testing, including urine culture.
  - 🕮 Asymptomatic UTIs in SCI patients managed with an indwelling catheter or IC are generally not treated.
  - Exceptions include patients undergoing invasive procedures (cystoscopy, urodynamics), presence of VUR, and at times during pregnancy.
- 🕮 Note: Urea splitting organisms produce struvite calculi made of ammonium and magnesium phosphate.
- **Symptomatic UTIs**
  - Malodorous urine, cloudy urine, sediment in urine, increased frequency of urination, decreased urine output, fever, chills, nausea, malaise, LE spasms, lower abdominal discomfort, bladder spasms, burning sensation, urinary leakage or incontinence, hematuria, discharge at suprapubic catheter site or at the urethral meatus, or signs and symptoms of AD in those with injuries at or above T6
  - Significant bacteria colony count (defined as >100 CFU/mL)
  - Pyuria (defined as >10 WBC/hpf)
  - Treated as a complicated UTI in the setting of NLUTD (5–7 days of antibiotics)
  - In febrile UTI, clinicians may consider ordering upper tract imaging if patient does not respond appropriately to antibiotic therapy OR the patient is not up to date with routine upper tract imaging.
  - In recurrent UTIs, clinicians should evaluate the upper and lower urinary tracts with imaging and cystoscopy.
  - Consider addition of a probiotic.
  - Fluid intake is increased and catheterization frequency is increased to reduce bacterial concentration and remove the urine that serves as a culture medium for bacterial growth.

Prevention of Urinary Tract Infections

- 🕮 The use of prophylactic antibiotics to prevent UTIs after SCI is generally not supported.
- Adequate hydration is encouraged.
- Avoid bladder overdistension, which can lead to relative bladder wall ischemia and increase risk of UTI.
- 🕮 Some of the complications can be prevented by adequately draining the bladder at pressures below 40 cm H_2O, either by IC (in conjunction with the use of medications) or by timely surgical relief of outflow obstructions that would not otherwise respond to medications.
- Vitamin C supplementation and methenamine salts can be used as acidifying agents to discourage bacterial growth.

MOST COMMON URINARY TRACT COMPLICATIONS IN THE NEUROGENIC BLADDER

LOWER URINARY TRACT DYSFUNCTION

- 🕮 Irregular, thickened bladder wall and small diverticuli—earliest changes
- VUR: 10% to 30% of poorly managed bladders lead to pyelonephritis and renal stones.
- Hydronephrosis and hydroureters caused by outlet obstruction
- Overdistended, areflexic bladder
- Reduced bladder compliance
- Kidney and bladder stones

BLADDER CALCULI

- Second most common urologic complication after SCI
- Indwelling catheter is the major risk factor for forming recurrent bladder stones.
- UTI with urease producing bacteria also increases risk for bladder stones.
- Can lead to:
  - Recurrent catheter blockage
  - Bladder distension causing AD and/or UTI
  - Hematuria
- Options for treatment include irrigation with saline, cystoscopy with aspiration, litholapaxy, lithotripsy, open surgery, and dissolution with Renacidin®, a solution containing citric acid, glucono delta-lactone, and magnesium carbonate.
- More frequent catheter changes (every 1–2 weeks) can be helpful for prevention of recurrent bladder stones.

SEXUAL DYSFUNCTION AFTER SPINAL CORD INJURY

Male Sexual Act

- Male erectile and ejaculatory functions are complex physiologic activities that require interaction between vascular, nervous, and endocrine systems.
- Erections are controlled by the *parasympathetic* nervous system.
- Ejaculations are controlled by the *sympathetic* nervous system.

Erections

- Controlled by a *reflex arc* that is mediated in the sacral spinal cord and is modulated by higher brainstem, subcortical, and cortical centers
- Reflex arc afferent limb:
  - Involves somatic afferent fibers that originate in the genital region and travel through the pudendal nerve into the sacral spinal cord
- Reflex arc efferent limb:
  - Involves parasympathetic fibers that originate in the sacral spinal cord and travel through the cauda equina via S2–S4 nerve roots
  - Postganglionic parasympathetic fibers secrete nitric oxide, which causes:
    - Relaxation of smooth muscle of the corpus cavernosum
    - Increased blood flow to the penile arteries—vascular sinusoids of the penis become engorged with blood, resulting in an erection.

Ejaculation

- Signals the culmination of the male sexual act and is primarily controlled by the sympathetic nervous system
- Similar to sympathetic innervation to the bladder, nerve fibers originate in the thoracolumbar spinal cord (T11–L2) and travel through the hypogastric plexus. They innervate the vas deferens, seminal vesicles, and ejaculatory ducts.

Erectile Dysfunction

- Men with SCI may obtain reflexogenic or psychogenic erections.
- Often times, the quality of erection is inadequate for intercourse. As such, the erection can be augmented or induced.

Reflexogenic Erections

- Occur in >90% of men with complete and incomplete UMN lesions; up to 25% with complete LMN lesions
- This demonstrates the importance of sacral parasympathetic neurons in reflex erections.
- Can occur independently of conscious awareness and supraspinal input (mediated by paraspinal division of autonomic nervous system through S2–S4 roots)
- Can occur secondary to manual stimulation of the genital region (however, once stimulation has been removed, the erection may no longer be sustained)

Psychogenic Erections

- Involve supraspinal effects from erotic stimuli that result in cortical modulation of the sacral reflex arc
- Present in approximately 50% of men with incomplete UMN lesions and 25% with complete LMN lesions
- Not usually seen in patients with complete UMN lesions
- Erection is mediated by central origin and psychological activation center.
- As previously noted, erections are more likely with incomplete lesions.

Methods to Induce Erections

- Oral pharmacotherapy with phosphodiesterases (e.g., sildenafil, tadalafil, vardenafil):
  - Used with success in the SCI population with UMN lesions
  - Avoid in patients taking nitrates and monitor for hypotension.
  - Caution in patients at risk for AD.
- Intracorporeal injections with prostaglandin E1, alpha-blockers, or vasodilators:
  - Counsel about risk of priapism.
- Penile implants:
  - Effective but have high failure rates
  - Risk of infection, penile erosion
- Penile vacuum devices
- Penile ring devices

Ejaculatory Dysfunction

- In men with SCI, the ability to ejaculate is less than the ability to obtain an erection.
- 🕮 The rate of ejaculation varies depending on the location and nature of the neurologic injury:
  - Approximately 5% to 15% of men with complete UMN lesions and 18% with complete LMN lesions have ejaculations.
  - The percentages are higher with incomplete injuries.
- Achieving ejaculation does not ensure successful reproduction, as sperm quality and motility are affected in SCI. An evaluation from a reproductive specialist may be needed.
- Semen analysis in men with SCI reveals decreased sperm count and sperm motility.
- Sperm retrieval in men who are unable to ejaculate:
  1. Penile vibratory stimulation (PVS):
     - Can be used at home
     - Caution in patients at risk for AD.
  2. Electroejaculation, if PVS unsuccessful:
     - May be painful in incomplete lesions
     - Caution in patients at risk for AD.
     - 🕮 Medical supervision required

3. Prostate massage
4. Surgical sperm removal. This can include testicular sperm extraction, aspiration, microsurgical epididymal sperm aspiration, percutaneous epididymal sperm aspiration, and aspiration of the sperm from the vas deferens.

Male Fertility After Spinal Cord Injury

- Fertility in men after SCI is impaired. As previously mentioned, two major causes are ejaculatory dysfunction and poor semen quality.
- 🕮 Poor semen quality is secondary to:
  - Stasis of prostatic fluid
  - Testicular hyperthermia
  - Recurrent UTIs
  - Abnormal testicular histology
  - Changes in hypothalamic–pituitary–testicular axis
  - Possible sperm antibodies
  - Type of bladder management
  - Long-term use of various medications

Prostatic Fluid Stasis

- Decreases sperm motility
- Studies have shown improvements in semen quality after 2 to 4 electroejaculations.

Sperm Counts and Motility Indices

- Sperm counts are lower in men with prostatic inflammation.
- Leukocytes (WBC $>10^6$) in the spermatic fluid reduced total sperm count, sperm velocity, and total motile sperm:
  - Single worst predictive factor for inability to penetrate an ovum is leukocyte concentration in the semen.
- Post-infection changes (testicular atrophy, epididymal duct obstruction) may affect fertility.

Abnormal Testicular Histology

- The most common finding noted on biopsy is atrophy of the seminiferous tubules.
- No investigations have found a significant correlation among biopsy findings, level of injury, length of injury, hormonal changes, or number of UTIs.

Female Sexual Act

- Sexual excitation with vaginal vasocongestion and lubrication is the result of psychogenic and physical stimulation.
- 🕮 Stimulation of the genital region, including clitoris, labia majora, and labia minora, causes afferent signals to travel via the pudendal nerve into the S2–S4 segments for reflexogenic vaginal vasocongestion.
- These fibers interact with the efferent parasympathetic fibers that project through the pelvic nerve, resulting in:
  - Dilation of arteries to perineal muscles and tightening of the introitus
  - Bartholin's glands secrete mucus, which aids in vaginal lubrication
- 🕮 Vasocongestion and lubrication are also related to sacral reflex preservation.
  - Reflexogenic
- If a *hyperactive bulbocavernosus* (BC) reflex is present, reflexogenic lubrication is usually possible, although quality of arousal might differ from preinjury levels.
- If a *hypoactive BC* reflex with some preservation of sensation within the S4–S5 dermatomes, reflexogenic lubrication is usually possible.
  - If BC reflex is absent and no sensation within S4–S5 dermatomes, then ability to have reflexogenic vaginal lubrication is lost.

  - Psychogenic
    - Ability to generate lubrication psychogenically is related to T11–L2 sensory preservation
  - If the BC and anocutaneous reflex are absent, and no sensation at S4–S5 is present, then attainment of orgasm is unlikely.
- Female orgasm is characterized by the rhythmic contraction of the pelvic structures. Female orgasm also results in cervical dilation, which may aid in sperm transport and fertility. Approximately 50% of SCI patients are able to achieve orgasm.
- Patients who have preserved sensation at T11–L2 dermatomes can achieve psychogenic vaginal lubrication.
- Sildenafil may improve subjective arousal when in conjunction with manual stimulation.
- Patients with UMN injuries can achieve reflexogenic vasocongestion by manual genital stimulation.
- LMN injuries at S2–S5 have reduced ability to achieve orgasm.
- 🕮 Decreased libido is reported after SCI and is likely due to a combination of psychological and physical changes after injury, including a change in self-image and altered sensation in the genital region.

Female Fertility After Spinal Cord Injury

- Immediately following SCI, amenorrhea occurs in 85% of women with cervical and high thoracic injuries and 50% to 60% of women overall.
- However, 50% and 90%, respectively, have return of menstruation within 6 to 12 months after injury.
- 🕮 SCI does not affect female fertility once menses return.

Birth Control

- Can be of concern for women with SCI. Consideration of not using early after injury, especially in patients at risk for thromboembolic disease.
- Condoms—provide protection
- Diaphragm—needs adequate hand dexterity
- Oral contraceptives—are associated with increased risk of thromboembolism
  - Consider progestogen-only oral contraceptives, or use combination pills that have low-dose estrogen. Implants can be used.
- The use of estrogen or depot medroxyprogesterone acetate injections should be avoided within the first year of injury and also in any women who smoke or who have a history of migraine, past history of DVT, or cardiovascular concerns.
- Intrauterine device—can increase risk of pelvic inflammatory disease, which can cause AD.

Pregnancy

- 🕮 The likelihood of pregnancy after SCI is unchanged because fertility is unimpaired.
- Pregnant women with SCI may develop:
  - Pressure injuries
  - Recurrent UTIs:
    - Not enough evidence to support prophylactic antibiotics for all pregnant patients with SCI
    - Frequent surveillance cultures and alterations in bladder management to decrease residual volumes are recommended.
  - Increased spasticity
  - Decreased pulmonary function
  - 🕮 AD may be the only clinical manifestation of labor:
    - Uterine innervation arises from T10–T12 level. Patients with lesions above T10 may not be able to perceive uterine contractions.
    - During labor and delivery, the risk of AD is 85% to 90% of patients with lesions at T6 and above.

- Treatment of choice is epidural anesthesia extending to T10 level.
- Epidural meperidine, bupivacaine, and fentanyl with bupivacaine have been effective in controlling labor-induced AD.
- Depth of general anesthesia needed to control labor-induced AD can induce neonatal depression and uterine atony.
- Avoid general anesthesia with depolarizing agents if the patient had SCI or other significant trauma within 1 year as it can cause hyperkalemia.
- Epidural should continue for at least 12 hours after the delivery or until AD resolves.
- If AD is refractory to epidural and regional anesthesia, urgent cesarean or operative vaginal delivery may be necessary.
- Need to distinguish AD from preeclampsia:
  - AD:
    - Severe headache and increased BP occur in synchrony with uterine contractions. BP and other symptoms normalize during relaxation of the uterus.
  - Preeclampsia:
    - High BP is more persistent in an individual with preeclampsia.
    - Preeclamptic patients also show proteinuria (>300 mg/dL), decreased platelets, and elevation in uric acid level and liver function tests, not present in AD.

– Slightly increased incidence of preterm labor and small birth weight infants
– Fetal hypoxemia can occur due to vasoconstriction of uterus/placenta:
  - Patients should be checked for cervical dilatation and effacement 1 to 2× weekly after 28 weeks' gestation.
  - Voluntary hospital admission could be offered after 36 weeks for close monitoring.
– Constipation
– Thromboembolism: Increased risk in acute SCI (first 6 months). Following this, the risk is almost the same as the general population:
  - Growing fetus increases pressure on the venous return from the legs.
  - Hypercoagulable state due to pregnancy-increased Factors I, VII, VIII, IX, and X; increased platelet activation, and decreased fibrinolytic activity
  - Increased risk due to decreased mobility
  - Loss of sympathetic input to vasoconstrict blood vessels
  - Symptoms: Leg edema, which may be unilateral; low-grade fever; area of the skin noticeably warmer than the general skin temperature; pain (in incomplete injuries)
  - Prevention:
    - Range of motion (ROM) exercises and changes in positioning to improve blood flow
    - Elevate lower limbs.
    - Use sequential compression devices, or wear gradient elastic compression stockings.
  - Insufficient evidence to recommend universal thromboprophylaxis during pregnancy in patients with preexisting SCI
  - If acute SCI should occur during pregnancy, the patient should receive chemical thromboprophylaxis as described for nonpregnant individuals:
    - Treatment for a DVT is similar to that for patients without SCI. Anticoagulation is continued for 6 months.
    - Low-molecular-weight heparin (LMWH) does not cross the placenta, is compatible with breastfeeding, and can be resumed hours after delivery.

GASTROINTESTINAL COMPLICATIONS AND BOWEL MANAGEMENT IN SPINAL CORD INJURY

Gastrointestinal Anatomy and Neuroregulatory Control (Figure 7–28)

- The colon is a closed tube bound proximally by the ileocecal valve and distally by the anal sphincter. It is composed of smooth muscle oriented in an inner circular and outer longitudinal layer.

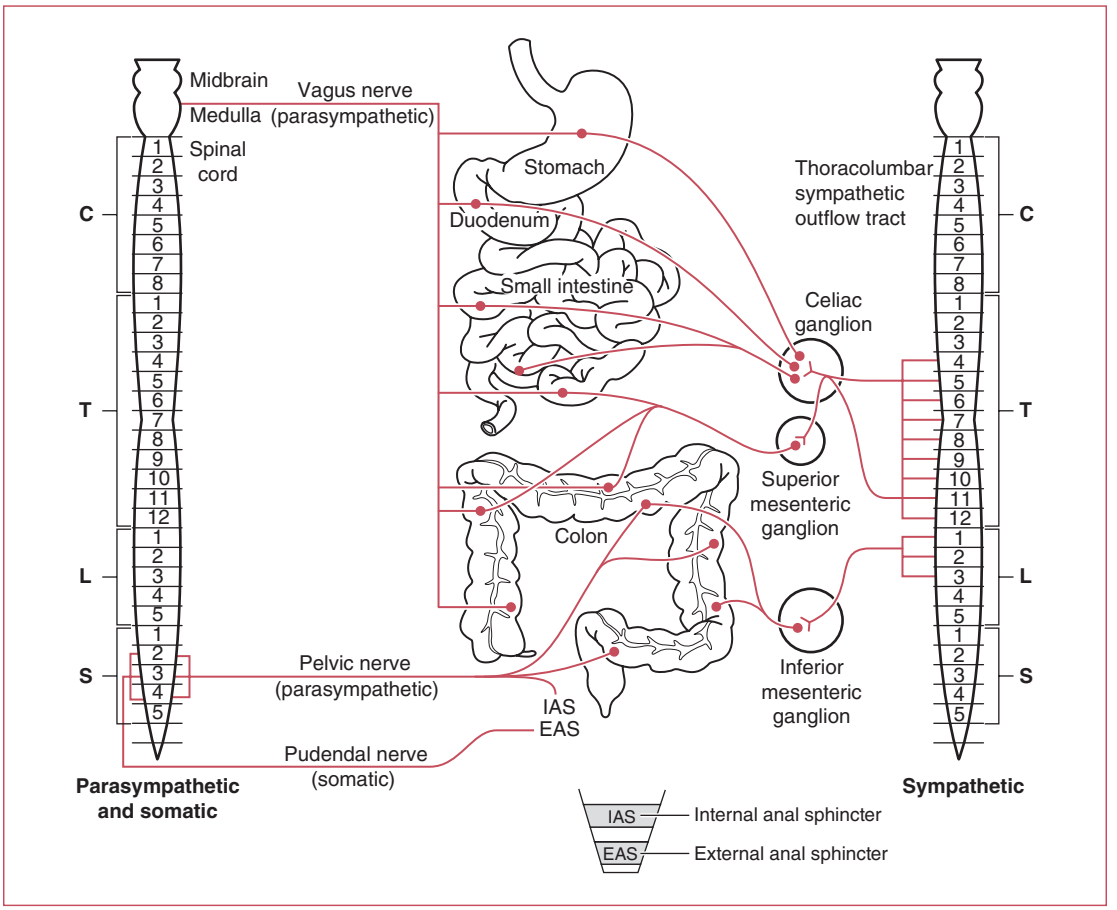


FIGURE 7–28 Innervation of the gut.
Source: Adapted from Nesathurai S, ed. *The Rehabilitation of People with Spinal Cord Injury: A House Officer's Guide*. 3rd ed. Arbuckle Publishers; 2013.

- The lower colon and anorectal region receive innervation by autonomic (sympathetic, parasympathetic) and somatic pathways.
- The intrinsic enteric nervous system (ENS) is composed of Auerbach's (myenteric; primarily motor) and Meissner's (submucosal; primarily sensory) plexi and coordinates the function of each segment of the bowel. They both lie between the walls of smooth muscle mentioned earlier.
- The parasympathetic and sympathetic nervous systems modulate the activity of the ENS, which in turn inhibits the inherent automaticity of the bowel's smooth muscle.

PARASYMPATHETIC NERVOUS SYSTEM

- Increases upper gastrointestinal (GI) tract motility
- Enhances colonic motility
- Stimulation provided by the action of the vagus nerve (innervates proximal to mid-transverse colon) and by the splanchnic nerves (pelvic nerve), which originate from the S2–S4 region and innervate the descending colon and rectal region

SYMPATHETIC NERVOUS SYSTEM (T12–L2)

- Inhibits colonic contractions
- Favors function of storage

- Innervation projects through the hypogastric nerve via superior mesenteric, inferior mesenteric, and celiac ganglia

SOMATIC NERVOUS SYSTEM

- Increases external anal sphincter (EAS) tone, promoting continence
- The EAS consists of a circular band of striated muscle that is part of the pelvic floor.

ANAL REGION

INTERNAL ANAL SPHINCTER

- Composed of smooth muscle under the influence of the sympathetic and parasympathetic systems. Sympathetic tone causes contraction of the internal anal sphincter, while parasympathetic tone causes relaxation of sphincter tone.
- Surrounds the anus proximally
- Contracted most of the time; relaxes with filling of the rectum in a neurologically intact individual

EXTERNAL ANAL SPHINCTER

- Composed of circular band of striated skeletal muscle and is part of the pelvic floor
- Helps to maintain continence by increasing its tone
- Innervated by somatic innervation from the pudendal nerve (S2–S4), which gives volitional control, learned by maturation and reflex activity
- Higher cortical centers and pontine defecation center send stimulus for EAS relaxation, allowing defecation.

STORAGE AND DEFECATION IN THE NEUROLOGICALLY INTACT INDIVIDUAL

STORAGE

- The internal anal sphincter is sympathetically activated (T11–L2), allowing for relaxation with filling of the rectum with stool bolus.
- EAS tone increases secondary to spinal cord reflexes and modulated action of higher cortical regions, maintaining continence.

DEFECATION

- Rectosigmoid distention causes reflex internal anal sphincter relaxation.
- Volitional cortical activity sends signal to pontine defecation center.
- Volitional contraction of the levator ani muscle allows opening of the proximal canal, relaxing the EAS and puborectalis muscles.
- Reflexive rectal propulsive contractions take place, resulting in expulsion of the stool bolus.

COLONIC DYSFUNCTION IN THE SPINAL CORD INJURY PATIENT

HYPERREFLEXIC (OFTEN CALLED CONAL OR UPPER MOTOR NEURON) NEUROGENIC BOWEL

- 🕮 In a UMN lesion such as an SCI, the GI system can be affected by loss of sympathetic and parasympathetic input at the transverse and descending colon, resulting in decreased fecal movement. Fecal impaction and constipation are the most common complications during recovery.
- Cortical control is disrupted with decreased ability to sense the urge to defecate.
- EAS cannot be voluntarily relaxed with such injuries, and pelvic floor muscles become spastic.
- However, nerve connections between the spinal cord and colon, as well as Auerbach's (myenteric) plexus, remain intact. Stool can be propelled by reflex activity.
- Possible decreased propulsion in distal colon

AREFLEXIC (OFTEN CALLED INFRACONAL OR LOWER MOTOR NEURON) NEUROGENIC BOWEL

- Lesion below conus medullaris (i.e., cauda equina syndrome)
- Reflex defecation is absent.
- Auerbach's (myenteric) plexus coordinates the movement of stool. However, movement is slow.
- Overall, constipation results with incontinence due to flaccid EAS.

Management of Bowel Dysfunction in Spinal Cord Injury

ACUTE PHASE

ADYNAMIC ILEUS AND GASTRIC ATONY

- Occur in approximately half of SCI patients, resulting from spinal shock and reflex depression
- Adynamic ileus usually presents immediately following SCI (can be delayed 24–48 hours) and typically resolves within 1 week.
- Gastric atony may result in vomiting and aspiration.
- **Management:**
  - Nasogastric tube (NGT) suction to prevent GI dilation and respiratory compromise for persistent abdominal distention
  - IV fluids
  - Parenteral nutrition if >3 days
  - If longer, metoclopramide and/or erythromycin can be used to stimulate peristalsis if other interventions are unsuccessful (metoclopramide works primarily in the stomach).
  - Neostigmine for refractory cases of pseudo-obstruction

CHRONIC PHASE—PERIOD OF MONTHS TO YEARS

- **Colonic distension:** Problems with small bowel motility
- **Pseudo-obstruction:** No evidence of obstruction on radiographic studies
- Abdominal distention, nausea, vomiting, constipation
- **Secondary causes:** Electrolyte imbalance and medications (narcotics, anticholinergics)
- **Management:**
  - NGT suction if gastric atony
  - Remove constipating medications if possible.
  - Oral medications to promote stool propulsion (see next section)
  - Rectal medications (i.e., suppositories, enemas)
  - If cecum is dilated >12 cm, consider GI and surgical evaluation.

LONG-TERM MANAGEMENT

- Padded upright commode preferred to side-lying position. If performed lying down, best to be in left lateral decubitus position.
- Maintain adequate fluid intake (2–3 L/day but need to coordinate with bladder management).
- Minimize medications that decrease bowel motility (opioids, tricyclic agents, anticholinergics) as much as possible.
- Diet: Moderate fiber intake (approximately 15–20 g/day)
- Medications:
  - Bowel stimulants and irritants
  - Stool softeners
  - Suppositories

Bowel Medications

- Stool softeners, such as docusate sodium, increase fat and fluid accumulation in the GI tract.
- Oral stimulants such as senna stimulate peristalsis by acting on Auerbach's plexus.
  - Additional oral medication can be used including polyethylene glycol (Miralax®).
- Bulk-forming agents promote evacuation by retaining or pulling H_2O into colon:
  - More often used in LMN bowel and not usually used in UMN bowel program initially
  - Examples: Psyllium (Metamucil®, Perdiem®, Konsyl®) or methylcellulose (Citrucel®)
- Suppositories should be placed high against the rectal wall:
  - Glycerin: Draws water into stool and stretches rectal wall
  - Bisacodyl (Dulcolax®) is in an oil base that stimulates peristalsis and sensory nerve endings.
  - Magic Bullet® is in a water base and acts faster than the Bisacodyl in an oil base.
  - Enemeez is a 5-mL mini-enema of docusate sodium. Enemeez-plus has added benzocaine.

Bowel Program

GOALS

- Frequency of bowel program depends on the person's premorbid habits, personal preference, and minimizing complications.
- Ultimate goal is consistent and complete evacuation of the bowel at a specified time, in a relatively short time period, without incontinence between programs.
- To assist with defecation, intact reflexes can be utilized:
- 🕮 **Gastrocolic reflex:**
  - Contraction of the colon occurring with gastric distention
  - When feasible, SCI patients should be instructed to perform their bowel programs 20 to 30 minutes after a meal. Increased colonic activity occurs in the first 30 to 60 minutes after a meal (usually within 15 minutes).
- 🕮 **Rectocolic reflex:**
  - Occurs when rectal contents stretch the bowel wall reflexively, relaxing the internal anal sphincter and leading to left colonic contraction
  - Suppositories and digital stimulation cause the bowel wall to stretch and take advantage of this reflex.
  - Note this reflex can be manipulated by *digital stimulation* of the rectum.
  - *Digital stimulation* is accomplished by gently inserting a gloved, lubricated finger into the rectum, and slowly rotating the finger in a clockwise circular motion until relaxation of the bowel wall is felt or stool/flatus passes (approximately 1 minute).
- 🕮 Key components of a bowel program depend on type of neurogenic bowel (hyperreflexive vs. areflexic bowel)
  - Hyperreflexive bowel program:
    - For pharmacologic intervention, one can start with a combination of stool softeners and stimulants with a suppository in what has been termed a "3-2-1" program:
      1. Stool softener (e.g., Colace®: 100 mg 3× daily)
      2. Oral stimulant (e.g., Senokot®: Two tablets daily, timed approximately 8 hours before suppository)
      3. Suppository (e.g., Dulcolax®—one suppository daily after meal—usually dinner or breakfast)
  - Others will start the use of polyethylene glycol orally instead of oral stool softeners and stimulants and then utilize a suppository.
    - Dietary fiber intake should also be encouraged.
    - Digital stimulation—insertion of gloved finger and move in circular motion to allow for EAS to relax
  - Areflexive bowel program:
    - Patient may not respond to suppositories.
    - Goal is firm formed stool that can pause manually with ease and does not pass accidentally between bowel care.
    - Use of bulking agents and softeners to obtain optimal consistency
    - Manual evacuation of stool 2 to 3×daily

Complications of Neurogenic Bowel

- Fecal incontinence can lead to skin breakdown and UTI.
- Fecal impaction: Nausea, abdominal discomfort, and AD. Some use lidocaine gel during disimpaction to avoid causing AD. Fecal impaction is the most common complication after the first month of injury.
- Anticholinergic meds that are prescribed for failure-to-store bladder, as well as opioid medications, can cause constipation.
- Bowel dysfunction affects the patient's community integration—socially, vocationally, and psychologically. If bowel dysfunction affects a person's quality of life (e.g., takes a long time, >1 hour, episodes of incontinence), then additional interventions should be considered.

- Transanal irrigation has been shown to be effective in such circumstances. Appropriate training is required.
- Surgical intervention, including anterograde continence enema (Malone procedure) and colostomy (or less frequently ileostomy), may also be considered if diet changes, medications, and the previously mentioned techniques fail to produce consistent bowel movements.

Other Gastrointestinal Complications in Spinal Cord Injury

GASTROESOPHAGEAL REFLUX

- Avoid prolonged recumbency; elevate the head of the bed.
- Avoid smoking.
- Avoid certain medications, such as Ca+ channel blockers, benzodiazepines, nitrates, and anticholinergics.
- Treatment:
  - Antacids for mild to moderate symptoms
  - H2 blockers, metoclopramide 10 mg four times a day (short-term use only because of side effects)

GASTROINTESTINAL BLEEDING

- Most frequently secondary to perforating and bleeding ulcers
- Most common within a few days of injury and peak during the first few weeks to first month after injury
- Increased risk on higher level injuries and neurologically complete
- Use of steroids may increase risk.
- Diagnosis: Endoscopy is the diagnostic method of choice.
- Provide prophylaxis for short term only (usually up to 1 month) unless otherwise indicated after SCI with H2 blockers, proton pump inhibitor, or sucralfate.
- Treatment:
  - With active GI bleeding: Maintain BP, correct coagulation deficits, monitor CBC, and consult GI/surgical service as appropriate.

CHOLECYSTITIS

- Most common cause of emergency abdominal surgery in chronic SCI patients
- 3× greater risk in SCI
- Possible causes: Abnormal gallbladder motility in lesions above T10, abnormal biliary secretion, abnormal enterohepatic circulation
- Should be considered if adynamic ileus does not resolve or it recurs
- Treatment:
  - Medical observation/antibiotics
  - May opt for surgical removal

PANCREATITIS

- Most common in the first month postinjury
- May be related to steroid use—increased viscosity of pancreatic secretions
- May suspect when adynamic ileus does not improve
- Clinical symptoms:
  - Abdominal pain
  - Nausea
  - Emesis
  - Poor appetite
- Evaluation:
  - Labs: Elevated amylase, lipase
  - Radiographs
  - CT of abdomen
  - Abdominal US

SUPERIOR MESENTERIC ARTERY SYNDROME (FIGURE 7–29)

- Superior mesenteric artery (SMA) syndrome is a rare condition in which the third portion of the duodenum, which is located between the aorta and SMA, is intermittently compressed by overlying SMA, resulting in GI obstruction.
- Predisposing factors include:
  - Rapid weight loss (decrease in protective mesenteric fat pad)
  - Prolonged supine position. Most common in tetraplegia.
  - Spinal orthosis
  - Flaccid abdominal wall causes hyperextension of the back.
- Symptoms:
  - Postprandial nausea and vomiting
  - Bloating
  - Abdominal pain
- Diagnosis: Upper gastrointestinal (UGI) series demonstrates abrupt duodenal obstruction to barium flow.
- Treatment is typically conservative:
  - Eat small, frequent meals in an upright position.
  - Lie in the left lateral decubitus position after eating.
  - Metoclopramide (Reglan®): Stimulates motility of upper GI tract (primarily stomach)
  - Rarely requires surgery. If conservative treatment fails, surgical duodenojejunostomy should be performed.
- *Remember:* Any condition that decreases the normal distance between the SMA and aorta (weight loss, supine position, Halo, flaccid abdominal wall) may result in compression of the duodenum, described as the nutcracker effect.

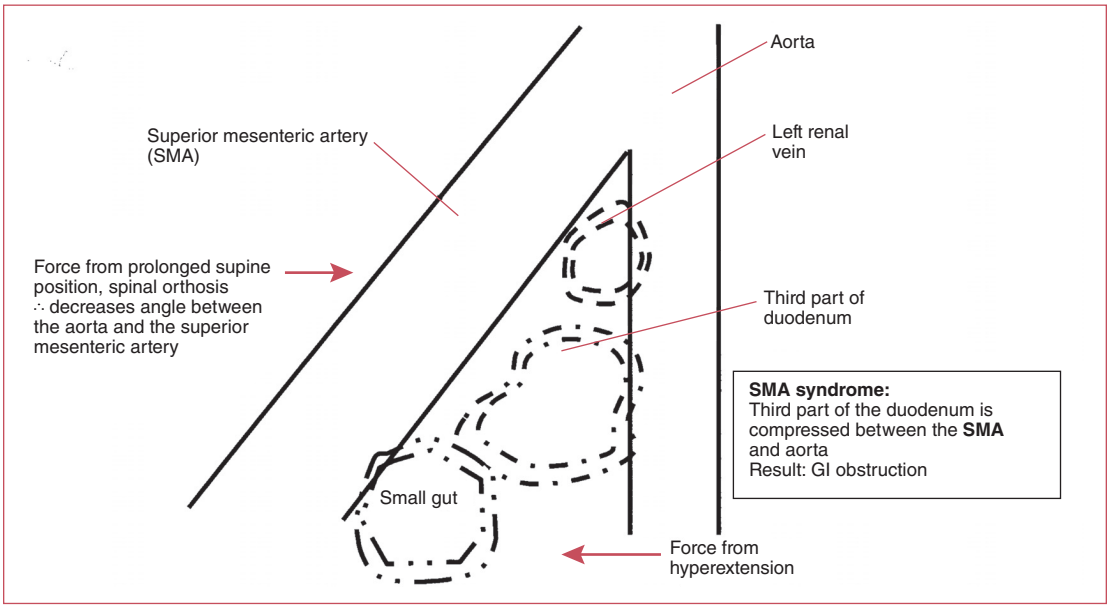


FIGURE 7–29 Lateral view through the duodenum and left renal vein.
GI, gastrointestinal.

METABOLIC COMPLICATIONS IN SPINAL CORD INJURY

Hypercalciuria

- Immobilization and reduced weight bearing result in uncoupling of the normal mechanism responsible for maintaining bone, promoting bone resorption and hypercalciuria.
- Vitamin D and parathyroid hormone are not involved in the process.

Hypercalcemia

- Incidence: 10% to 23% of persons with SCI
- More common in adolescent and young adult males
- More common in patients with tetraplegia than with paraplegia
- 🕮 Usually appears 4 to 8 weeks after SCI (2 weeks–6 months postinjury)
- Clinical presentation: Often insidious and presenting symptoms can be vague. Should maintain a high index of suspicion.
- Signs and symptoms: "Bones, stones, and psychiatric overtones":
  - Fatigue
  - Lethargy
  - Dehydration
  - Constipation
  - Anorexia nausea
  - Vomiting
  - Polydipsia
  - Polyuria
  - Psychosis
- Labs: Serum calcium level is usually <10.5. Since serum calcium is protein bound, the serum calcium must be corrected for albumin concentration.
- Treatment:
  - Treat both symptomatic and asymptomatic patients because prolonged hypercalcemia can cause nephrocalcinosis.
  - 🕮 IV fluid hydration with normal saline. Increases urinary excretion of calcium.
  - Early mobilization
  - Diet restrictions:
    - Restriction of dietary calcium intake is unnecessary; 1, 25-dihydroxy vitamin D levels already are low, suppressing intestinal absorption of calcium.
    - Restriction of intake of vitamin C may be prudent.
  - Medications:
    - Pamidronate: Inhibits osteoclast-mediated resorption and reduces osteoclast viability. The drug is administered as a single IV infusion (60 mg × 4 hours to 90 mg × 24 hours, depending on severity of hypercalcemia) and rapidly lowers serum calcium within 3 days. The serum calcium falls to a nadir within 7 days and may remain normal for several weeks or longer. Can be repeated if needed. The IV fluids can be discontinued 2 to 3 days after pamidronate is completed.
    - Furosemide increases urinary excretion of calcium (although not routinely used). Avoid thiazide diuretics → cause hypercalcemic effects.
    - Other meds include calcitonin.
  - Check serial labs.

Osteoporosis (Consortium for Spinal Cord Medicine 2022)

- Rapid bone loss occurs below the level of injury and primarily at load-bearing sites.
- On average, bone mineral density losses at the hip decrease by approximately 2% per month in acute motor complete SCI.
- Etiology is likely multifactorial:
  - Mechanical unloading (i.e., lack of weight bearing) is thought to play a major role.
  - Neural, hormonal, vascular, autoimmune, nutritional, and behavioral factors may also be involved.
- Secondary causes of osteoporosis, such as thyroid disease, parathyroid disease, hypogonadism, renal disease, and chronic liver disease, have been shown to be present in approximately 30% of patients with SCI and should be considered.

- Long-term use of certain medications such as heparin, anticonvulsants, and opioids may also contribute to bone loss.
- Laboratory screening:
  - Serum 25-hydroxyvitamin D (25-(OH)D)—checked annually and 12 weeks following repletion
  - CBC
  - Ionized calcium
  - Phosphate
  - Parathyroid hormone (PTH)
  - Creatinine
  - Bone-specific alkaline phosphatase and transaminases
  - Hemoglobin A1c
  - Thyroid stimulating hormone (TSH)
- Bone density testing:
  - The Consortium Guidelines has recommended that adults with SCI resulting in permanent motor or sensory dysfunction should have dual-energy x-ray absorptiometry (DXA) scan of total hip, proximal tibia, and distal femur as soon as medically stable with follow-up at intervals of 1 to 2 years
- Treatment:
  - Vitamin D supplementation is recommended with a maintenance dose of vitamin D3 (1,000–2,000 IU/day).
  - Calcium supplementation is dependent on age and sex (not appropriate if individuals are hypercalcemic) and usually approximately 1,000 mg/day.
  - Therapy-based treatments include passive standing for 1 hour 5× per week and/or suggested functional electrical stimulation (FES) or neuromuscular electrical stimulation (NMES). However, there are limited data to show that passive standing will have any effect and that NMES would need to be used consistently to have some effect in preventing bone loss.
  - Recommended to have risk/benefit discussion with patient for use of medications including for prevention of and/or treatment for osteoporosis including the use of bisphosphonates, zoledronic acid, or denosumab.

Fractures

- Fractures occur in 25% to 50% of individuals with chronic SCI:
  - Knee (distal femur and proximal tibia) is the most common site.
  - The average time to fracture is approximately 9 years postinjury.
- Most fragility fractures (those that occur specifically due to low bone mineral in persons with chronic SCI) occur while performing activities of daily living (ADLs).
- Fracture-related complications (e.g., nonunion, contracture, skin breakdown, DVT) occur in over 50% of cases.
- Risk factors for fracture include:
  - Prior fragility fracture (fracture caused by force insufficient to fracture normal bone)
  - Family history of fracture
  - Female gender
  - Age (bimodal: ≤16 years old and increasing age)
  - Time postinjury (>10 years)
  - Body mass index <19
  - Paraplegia
  - Motor complete SCI
  - Alcohol intake >5 servings per day
- Diagnosis: Workup with plain x-ray is usually sufficient.
- Treatment:

- The main goals of treatment in chronic SCI for persons who do not use their LEs for mobility are to minimize complications and to preserve pre-fracture function.
- 🕮 Fractures should be evaluated by orthopedic consultation. LE fractures above the knee should be managed operatively, while fractures below the knee may consider operative intervention or conservative management. Nonoperative treatment consists of soft padded splints (e.g., a well-padded knee immobilizer for femoral supracondylar, femoral shaft, and proximal tibial fractures; and a well-padded ankle immobilizer for distal tibial fractures).
- With nonoperative management, the patient is typically allowed to sit within a few days. Callus formation is usually evident at 3 to 4 weeks. However, ROM is initiated at 6 to 8 weeks, with weight bearing delayed for a longer period.
- Newer evidence suggests that the use of surgery to treat fractures after SCI is increasing and that in most cases outcomes are good.

🕮 Cardiovascular Disease in Spinal Cord Injury

- In SCI, cardiovascular disease occurs more frequently and at an earlier age.
- Reduced energy expenditure is rarely compensated with a concomitant reduction in energy intake, often leading to morbid obesity.
- Adipose-derived proinflammatory cytokines, adipokines, prothrombotic agents, and elevated nonesterified fatty acids result in accelerated atherogenesis and ischemic heart disease in persons with SCI.
- Cardiovascular disease is a leading cause of mortality among people with SCI, accounting for between 18.4% and 25% of deaths.
- Increased risk factors in SCI:
  - Low high-density lipoprotein (HDL)
  - High total cholesterol and low-density lipoprotein (LDL)
  - Increased inflammatory markers: C-reactive protein (CRP) level high
  - Decreased fat-free body mass, increased obesity and visceral adipose tissue
  - Increased rate of smoking
  - Physical inactivity
  - Those with tetraplegia and neurologically complete injuries have a higher risk. Higher prevalence of insulin resistance, diabetes, metabolic syndrome.
  - Screening: HDL, LDL, total cholesterol, BP
- Nutritional assessment of diet; evaluate for obesity
- Evaluate activity level.
- Management:
  - 🕮 Primary goal is lowering LDL.
- For adults aged ≥20 years, fasting lipoprotein profile (total cholesterol, LDL, HDL, triglyceride [TG]) should be obtained every 5 years.
- If testing is nonfasting, then only *total cholesterol* and HDL are usable.
- If total cholesterol ≥200 mg/dL or HDL ≤40 mg/dL, then repeat the test to determine appropriate management.

Hyperglycemia and Metabolic Syndrome

- Up to 70% of the patients show insulin resistance (abnormal response to glucose load).
- Important to monitor in chronic patient and initiate treatment when appropriate
- Metabolic syndrome disturbances of carbohydrate and lipid metabolism, most commonly in men with SCI, are common and usually characterized by central obesity, abnormal carbohydrate metabolism, elevated BP, high triglycerides, high LDL, and low HDL values.
- Interventions:
  - Smoking cessation
  - Weight loss
  - Increase activity levels: Exercise 3× per week

  - Both FES and arm ergometry (high-intensity target HR 70%–80% of max HR predicted) have increased glucose tolerance and reduced lipid profile in SCI.
  - Modify diet.
  - Pharmacotherapy used if hypertension (HTN), hyperglycemia, and dyslipidemia do not respond to the previous actions

PULMONARY CARE AND RESPIRATORY COMPLICATIONS IN SPINAL CORD INJURY

General

- Respiratory complications occur in up to 67% of acutely injured patients.
- Pulmonary complications are more common in high cervical injuries (C1–C4) but occur in a significant portion of lower cervical and thoracic level injuries.
- 🕮 The *most frequent complications* are pneumonia, atelectasis, and ventilatory failure:
  - Respiratory complications are most common in the first year following injury but persist throughout life. Within the first 15 years following SCI, respiratory illnesses comprise 20% to 24% of all deaths.
  - The diaphragm (innervated by C3–C5) is the major muscle of inspiration contributing approximately 65% to the vital capacity (VC).
  - The inability to clear secretions in persons with SCI is a key factor in the development of pulmonary complications such as atelectasis and pneumonia. The primary muscles of expiration are the abdominal muscles including the rectus abdominus, transversus abdominus, internal and external obliques (T4–L2), and the internal intercostal (II) muscles of the lower rib cage (T6–T12).
  - For persons with cervical SCI, the majority require assisted ventilation upon initial hospital admission, with reports ranging from 54% to 65%. The need is higher in patients with complete injuries and higher levels of cervical injury.
  - For persons with neurologically complete injuries with high cervical injuries, reported rates of successful weaning from mechanical ventilation (MV) are 0% at C1, 0% to 28% at C2, 25% to 50% at C3, and 77% to 83% at C4.
  - Most patients with lower cervical injuries (C5–C6) are eventually able to breathe comfortably without mechanical assistance due to preservation of diaphragm function.
- Respiratory ability based on SCI level:
  - C3–C4: Respiratory failure secondary to disruption of diaphragmatic innervation, usually requiring MV early after injury. Usually able to wean off the ventilator.
  - Injuries above C8: Loss of all abdominal and intercostal muscles, impairment of inspiration and expiration
  - T1 through T5: Intercostal volitional function is lost.
  - T5 through T12: Progressive loss of abdominal motor function, impairing forceful expiration or cough
- Pulmonary dysfunction occurs for several reasons following SCI:
  - Paralysis of some or all respiratory muscles to varying degrees
  - Loss of ability to cough secondary to varying levels of abdominal muscle paralysis
  - Injury to chest—for example, rib fracture
  - Pulmonary injury—for example, lung contusion
- Thoracic injuries can present with pleural effusion, atelectasis, pneumothorax, hemothorax, or all of these.

Predisposing Factors for Pulmonary Complications

- Older age
- Obesity—restrictive respiratory deficits
- History of chronic obstructive pulmonary disease, asthma, and smoking
- 🕮 Pneumonia is the leading cause of death among chronic SCI patients.

Lung Physiology

INSPIRATORY MUSCLES

- Diaphragm: Main respiratory muscle during quiet breathing (75% of volume change). Contracts during inspiration and relaxes during expiration.
- External intercostals
- Accessory muscles

EXPIRATORY MUSCLES

- Internal intercostals
- Abdominal muscles are primarily active during forceful expiration and coughing. They also help push the relaxed diaphragm toward the chest cavity.

INSPIRATION IN THE NORMAL LUNG/INSPIRATION IN A LUNG WITH INSULT TO THE PHRENIC NERVE (FIGURE 7–30)

Pulmonary Function in Spinal Cord Injury—Restrictive Respiratory Changes

- The estimated loss of forced vital capacity (FVC) is significant at approximately 70% of predicted in individuals with an acute neurologically complete injury between C5 and C6. This value improves over time. In those with incomplete injuries as well as lower cervical levels, FVC is significantly higher.
- With development of intercostal and abdominal spasticity, FVC *can improve to 50% to 60%* of the predicted normal value.
- For those with incomplete injuries as well as lower cervical levels, FVC is significantly higher.
- 🕮 Tetraplegics usually develop *restrictive lung patterns:*
  - All volumes shrink (except residual volume).
  - If VC <1 L: Consider ventilation mechanically.
  - Once VC is >15 to 20 mL/kg, the individual can usually wean off ventilator.

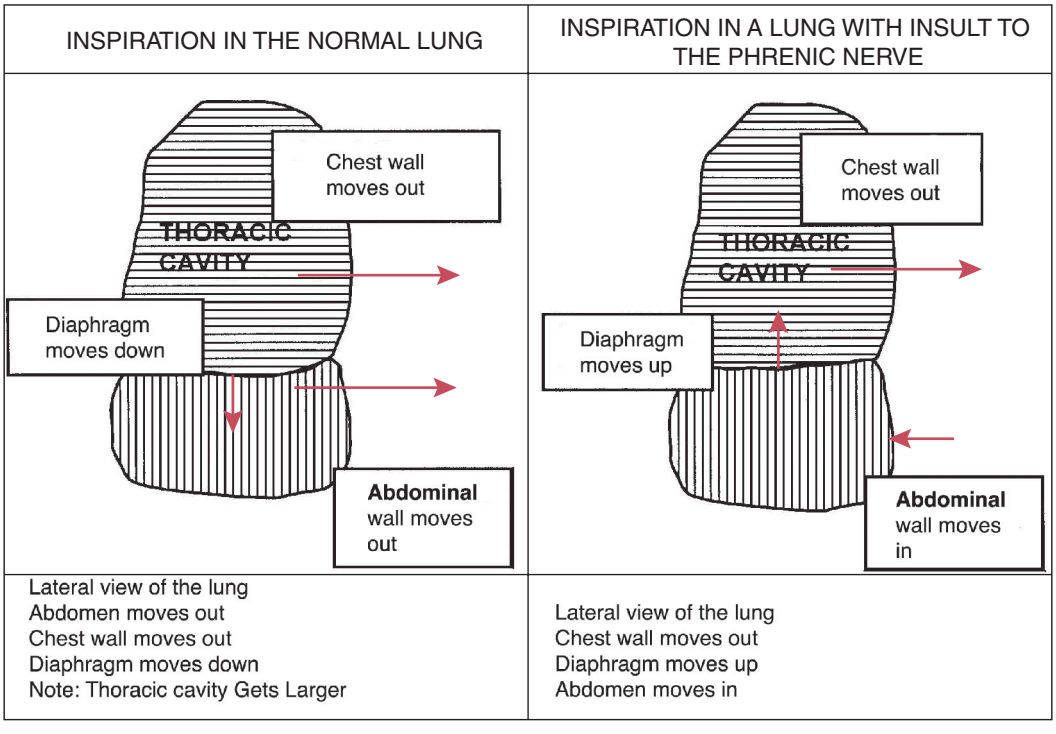


FIGURE 7–30 Inspiration in the normal lung and in a lung with insult to the phrenic nerve.

- Signs of impending respiratory failure:
  - Increased respiratory rate with decreased tidal volume
  - Decreased FVC <15 mL/kg body weight
  - Neurologic level C3 or higher
  - Patient cannot count to 15 slowly.
- 🕮 Mechanically ventilate when:
  - VC <1 L
  - Arterial blood gas (ABG) shows increasing PCO_2 or decreasing PO_2 levels.
  - PO_2 <50 mmHg
  - PCO_2 >50 mmHg
  - Severe atelectasis

Prevention and Treatment of Respiratory Complications

- Monitor VC.
- Assisted coughing techniques are beneficial. Manual cough assist ("quad cough")—placing the hands on each side of the patient's upper abdomen and applying intermittent pressure, coordinated with the initiation of cough by the patient—helps produce forceful cough.
- Suctioning—tracheal suctioning may cause increased vagal tone leading to bradycardia (only suction as you withdraw catheter). Suctioning does not adequately clear the left side of the lung due to the left main stem bronchus branching off at a 40- to 50-degree angle from vertical, making it more difficult to clear secretions.
- Mechanical cough assist—via use of an insufflation–exsufflation machine:
  - Provides higher peak cough flow than manual assisted cough
  - Advantages include the ability to better clear secretions, since it can clear both lung fields as well as clear out larger mucous plugs than the suction catheter, is easy to use, and patients prefer it.
  - Contraindicated in patients with a history of bullous emphysema, known susceptibility to pneumothorax or pneumo-mediastinum, or known to have had any recent barotrauma
- Chest physical therapy—see Chapter 9, Pulmonary, Cardiac, and Cancer Rehabilitation.
- Strengthening of pectoralis major muscle, clavicular portion, in tetraplegic patients is helpful.
- Glossopharyngeal breathing—stroking maneuver to force air into the lungs by the use of the lips, soft palate, mouth, tongue, pharynx, and larynx, followed by passive exhalation
- Pneumobelt—helps with exhalation. Inflatable, compresses the abdominal wall, diaphragm rises, and active respiration is produced.
- Inspiratory muscle training may be helpful.
- An abdominal binder can also be used to augment a cough effort.
- Use of bronchodilators is also an important adjunct of therapy and facilitates secretion management and reversal of atelectasis in patients with acute SCI.
- Compared with able-bodied individuals, VC is significantly higher in the supine compared with the upright posture in tetraplegic patients. In the supine posture, the abdominal contents displace the diaphragm cephalad causing it to lengthen, placing the diaphragm on a more favorable position on its length–tension curve. In the erect posture, the abdominal contents fall forward, causing the diaphragm to flatten and its fibers to shorten.
- Noninvasive means of ventilation (via nasal or full-face mask) may be employed in some specialty centers to prevent intubation for acutely injured patients.
- Weaning from ventilator in SCI:
  - Useful indices for weaning include maximum inspiratory pressure (MIP) of −20 cm H_2O or more negative and VC >10 to 15 mL/kg ideal weight.
  - The use of higher tidal volumes in the range of 15 to 20 mL/kg in patients with SCI had previously been recommended in persons with SCI, although time to weaning does not seem to be affected. This is less commonly being used today. If using higher tidal volumes, pressure plateaus should be maintained below 30 cm H_2O to prevent atelectasis, and the patient should not have adult respiratory distress syndrome.
  - The method with the highest success rates for weaning off MV is progressive free breathing, with or without the use of positive end-expiratory positive pressure (5 cm H_2O or less).

Pulmonary Compromise Related to Level of Injury

- Head trauma may disrupt respiratory drive.
- Lesions above C3 (and incomplete lesions initially)
  - Initially they require ventilatory support.
  - Later they will fall into two groups:
    1. No damage to phrenic nerve nucleus
    2. Damage to phrenic nucleus

EMG of diaphragm is important to rule out damage to the phrenic nerve nucleus if considering pacing for ventilation.

EMG, electromyography.

1. No damage to phrenic nerve nucleus
Lesion above the phrenic nerve nucleus innervation (C2 and above)
- Diaphragm is innervated by the phrenic nerve (C3–C5; nerve roots "C3, C4, C5 keep you alive").
- Lesion affecting the phrenic nerve can be determined by electrodiagnostic studies including nerve conduction of the phrenic nerve or EMG of the diaphragm.
- If phrenic nerve nuclei are intact and a UMN injury occurs, the patient may be a candidate for phrenic nerve or diaphragmatic pacing. See sections on Phrenic Nerve Pacing and Diaphragmatic Pacing.

2. Damage to the phrenic nerve
Lesions affecting C3–C5 nerve roots or phrenic nerve nucleus
- Determined by electrodiagnostic studies as previously noted
- If there is an LMN injury to the phrenic nerve, the patient is NOT a candidate for phrenic nerve pacing unless nerve grafts are utilized.

EMG, electromyography; LMN, lower motor neuron; UMN, upper motor neuron.

Pacing for Ventilation

- There are two types of pacing available, although both involve stimulating parts of the phrenic nerve: (a) phrenic nerve pacing and (b) direct pacing of the phrenic nerve on the diaphragm, often termed "diaphragmatic pacing."
- 🕮 **Pacing has several potential benefits compared with MV:** Improved quality of life, engagement of the patient's own breathing muscles, improved level of comfort, improved speech, restoration of olfactory sensation, increased mobility, reduced anxiety and embarrassment, elimination of ventilator noise, and reduced overall costs.
- In individuals with bilateral phrenic nerve function, full-time pacing is achieved in approximately 50% of cases and can be utilized as the sole mode of respiration. More than 80% to 90% of patients, however, can achieve significant freedom from MV (e.g., dependence on MV only at night).

A. PHRENIC NERVE PACING

- The technique involves electrical stimulation of intact phrenic nerves via surgically implanted electrodes to contract the diaphragm.
- Used successfully in patients with central hypoventilation and high tetraplegia
- **Contraindications to phrenic nerve pacing:**
  - Denervated diaphragm (determined in electromyography [EMG]), unless nerve grafting is considered
  - Significant lung impairment
- **Major complications of phrenic pacemaker:**
  - 🕮 *Signs of failure of pacemaker:*
    - Sharp chest pain
    - Shortness of breath
    - Absence of breath
    - Erratic pacing

  - *Causes of phrenic pacing failure:*
    - Diaphragmatic failure—due to overly aggressive pacing schedule
    - Infection of lung and/or phrenic nerve
    - Meds: Including sedatives, tranquilizers, and narcotics
    - Upper airway obstruction—tracheal aspiration
- **Phrenic motor study:**
  - Recording site: Diaphragm muscle
  - G1: Two finger breadths above the xiphoid process
  - G2: Anterior costal margin 16 cm from G1
  - Stimulation site: Lateral neck, posterior to the sternocleidomastoid (SCM) muscle, approximately 3 cm above the clavicle (Preston & Shapiro, 2013)

B. DIAPHRAGMATIC PACING

- Successful diaphragmatic pacing depends upon stimulation of the phrenic nerves via the intramuscular pathway.
- Less invasive procedure (laparoscopic) relative to phrenic nerve pacing
- This method involves wires emanating through the skin, whereas the entire stimulation system is implanted under the skin with the direct phrenic nerve stimulation approach.

Sleep Apnea in Spinal Cord Injury

- The incidence of obstructive sleep apnea in patients with SCI is significantly higher compared with the able-bodied.
- Sleep apnea occurs in approximately 15% to 60% of patients with acute and chronic SCI, with the higher numbers for mild symptoms.
- In SCI patients, sleep apnea is primarily obstructive, with a small percentage of patients demonstrating central sleep apnea.
- Risk factors:
  - Obesity, male, tetraplegia, neck circumference, and advancing age
  - Medications such as opiates and benzodiazepines as well as antispasticity drugs and antiarrhythmics may also significantly worsen sleep apnea.
- Common features include loud snoring, disrupted sleep, witnessed apnea, nocturnal gasping and choking, daytime sleepiness, morning headaches, and fatigue.
- Complications include daytime sleepiness and cognitive changes including poor attention, concentration, complex problem-solving, short-term recall, and judgment.
- In addition, there is increased risk for the development of HTN, pulmonary HTN, congestive heart failure, depression, and mortality.
- It is important to review with the SCI patient about sleep disturbance, snoring, and daytime somnolence:
  - Optimal workup would include an overnight oximetry recording, followed by a formal sleep study, if the oximetry recording is abnormal.
- Treatment includes assisted ventilation at night as well as possible use of medications directed toward relief of upper airway symptoms.

HETEROTOPIC OSSIFICATION

- Formation of mature lamellar bone indistinguishable from normal bone in soft tissues, most frequently deposited around a joint
- **Causes:**
  - No definitive pathogenesis established
  - It is unclear what the initiating factor is that results in the differentiation of mesenchymal cells into osteoblasts, which form new bone. Bone morphogenic protein has been implicated as well as soft tissue inflammation.

  - A decrease in tissue oxygenation or induced changes in multipotential connective tissue cells in which new bone forms in planes between connective tissue layers
- **Incidence:**
  - Heterotopic ossification (HO) has been reported to occur in 1% to 50% of patients following SCI from various reports in literature.
  - Clinically significant HO that results in significant limitation of joint ROM affects 10% to 20% of SCI patients; 5% to 8% progress to joint ankylosis.
  - Occurs below the level of neurologic injury
  - 🕮 Most common joints involved in SCI (in order of occurrence): Hip (anteromedial aspects), over 60%, followed by the knee, over 30%.
- 🕮 **Onset:**
  - The 1 to 3 months status postinjury is most common; peak at 2 months.
  - Can present after first 6 months. When it occurs after 1 year, it is usually associated with an acute fracture, DVT, or pressure injury. Usually has a more benign course.
- **Symptoms:**
  - Heat/warmth
  - Localized soft tissue swelling—may present similar to an acute DVT
  - Decreased ROM of a joint
  - Joint erythema/joint effusion
  - Low-grade fever
- **Risk factors:**
  - Spasticity
  - Completeness of injuries
  - Trauma or prior surgery to joint
  - Age: Younger age, although children have lower incidence
  - Pressure injury in proximity of joint
  - Presence of DVT
- **Diagnosis:**
  - Can be seen early after onset on triple-phase bone scan (seen early on the first two phases that reveal hyperemia and blood pooling; the third phase that reveals calcification is positive several weeks later). The bone scan returns to normal as HO matures 6 to 18 months postinjury.
  - 🕮 Bone scan is the most sensitive test for diagnosis and the earliest detection can occur 2.5 weeks post-trauma. Bone scan precedes findings on x-ray. Good to assess the maturity of the heterotopic bone.
  - Plain film detects HO at least 7 to 10 days after clinical signs are observed.
  - Serum alkaline phosphatase increases at 2 weeks, exceeds normal levels at 3 weeks, peaks at 10 weeks, and returns to normal by 18 weeks before HO matures. Not specific for HO and the absolute level of alkaline phosphatase does not correlate with the amount or degree of bone activity.
  - Creatine phosphokinase, CRP, and erythrocyte sedimentation rate can be elevated in acute HO but are not specific for HO.
  - A rapid increase in 24-hour prostaglandin E2 urinary secretion has been shown to suggest HO and can indicate the need for further imaging studies.
  - Ultrasonogram may be able to assist early in the development of HO.
  - MRI with increased T2 signal in soft tissues may assist in detection in the acute stage.
- **Complications:**
  - Decreased ROM/loss of function, which can lead to ankylosis
  - HO overlying a bony prominence will directly predispose to pressure injury/skin breakdown secondary to poor positioning.
  - Peripheral nerve entrapment
- **Prophylaxis:**
  - Studies have shown significantly lower incidence rate for those who received nonsteroidal anti-inflammatory drugs (NSAIDs) for HO prophylaxis. However, it is rarely used in SCI prophylaxis.

  - Radiation has been found to be effective for traumatic HO and hip surgery, however is not routinely used in the SCI population.
- **Treatment options**: No definitive standard treatment. Recommendations include the following:
  - ROM: The goal is to maintain functional range. Once the acute inflammatory period is over (i.e., warmth, erythema), the affected joint should be gently ranged through functional range. Vigorous force should not be used as this may lead to further ectopic bone formation. FES is a relative contraindication during the acute phase of HO.
  - Extracorporeal shock therapy can reduce pain and improve ROM.
    - In the presence of new HO, nonsteroidals (NSAIDs) can be used for treatment given that the previous mainstay of treatment in first-generation bisphosphonates (Didronel®) is no longer available. The most common interventions include indomethacin or celecoxib, the latter of which has the added benefit of less GI side effects.
  - Radiation therapy: Prevents further HO formation and prevents recurrence after surgery. Not often used acutely. Risks of use include delayed wound healing, risk of osteonecrosis, and sarcoma.
  - Surgery can be considered when HO severely limits ROM, impairing function. Usually recommended when bone is mature (12–18 months postinjury with normal bone scan). Various surgical approaches have been described, with wedge resection most commonly performed at the hip. While effective, there is high risk of infection.

DEEP VENOUS THROMBOSIS AND PULMONARY EMBOLISM IN SPINAL CORD INJURY

Deep Venous Thrombosis

- **Predisposing factors:**
  - Virchow's triad: Venous stasis/intimal injury/hypercoagulability
  - LE fractures
  - Obesity
  - History of previous DVT
  - Diabetes
  - Arterial vascular disease
  - Older age
  - Immobility
  - Malignancy
- **Incidence:**
  - 47% to 100% without prophylaxis; approximately 14% with appropriate chemoprophylaxis. Varies widely depending on the method of detection and number of cases evaluated in the study.
  - More common in neurologically complete patients
  - More common in patients with tetraplegia
- **Onset:**
  - Most common during first 2 weeks after SCI, with greatest risk of up to 8 weeks
  - Incidence decreases after 8 to 12 weeks post-SCI.
- **Diagnosis:**
  - 🕮 LE venogram is the gold standard.
  - D-dimer—high sensitivity but low specificity
  - Venous duplex can be used as a screening test for LE DVTs (better for proximal clots than distal).
- **Complications:**
  - 🕮 Pulmonary embolism (PE) is a leading cause of death during the first year after SCI (DeVivo, 2022).
  - Postphlebitic syndrome (late complication of DVT):
    - Distal venous HTN: Residual obstruction of outflow—incompetent valve
    - Swelling

- Spasticity
- Chronic pain (postthrombotic syndrome)
- AD

Pulmonary Embolism

- **Symptoms:**
  - Pleuritic chest pain
  - Dyspnea
  - Tachycardia
  - Fever
  - Hemoptysis
  - Hypoxemia
- **Physical examination:**
  - Increased S2 sound: Severe pulmonary HTN → cor pulmonale → right heart failure
  - Dullness at bases of lungs
- **Incidence:**
  - Up to 7% of acute/subacute SCI patients
  - Most cases develop from deep veins of the LEs.
  - PE is not influenced by degree or level of injury.
- **Diagnosis (PE):**
  - EKG: Right axis deviation; right bundle branch block (RBBB) if massive PE
  - ABG: ↓ PO_2 (PO_2 drops severely)
  - D-dimer—high sensitivity but low specificity
  - Chest x-ray:
    - Wedge-shaped opacity
    - Fluid
    - Vascularity
  - Perfusion lung scan: Ventilation/perfusion (V/Q) mismatched
  - Spiral (helical) CT scan
  - 🕮 *Gold standard:* Pulmonary arteriogram
  - MRI
- **Treatment (PE):**
  - Supplemental O_2 may be required
  - Anticoagulation: Heparin, LMWH, direct oral anticoagulation (DOACs)
  - Vasopressor to treat shock
- **Surgical treatment:**
  - Embolectomy

Prophylaxis for Thromboembolic Disorders

- **Mechanical prophylaxis:**
  - External pneumatic compression devices (effective) for the very acute time period
    - Improves LE venous return, reduces stasis, and stimulates fibrinolysis
    - If placement is delayed >72 hours, rule out DVT (venous duplex scan) before using compression devices
    - Not often used in rehabilitation as cannot keep on for 23 hours per day
    - Contraindicated in patients with arterial insufficiency
  - Electrical stimulation: Has been shown to be effective but hardly used
- **Chemoprophylaxis:**
  - Anticoagulants generally withheld for the first 72 hours. Incidence during that time period is small and perhaps increases complications. Can still use mechanical devices.
  - Hold dose morning of surgery and resume the next day.
  - The Consortium for Spinal Cord Medicine (2016) clinical practice guidelines recommend LMWH to be initiated as chemoprophylaxis once there is no sign of active bleeding in acute SCI.

  - A box warning exists regarding the risk of spinal or epidural hematoma formation in patients who receive LMWH. (Important to clear with surgeon when LMWH can be initiated if patient has undergone spinal surgery.)
  - DOACs are starting to be used; further study is underway.
- **Inferior vena cava (IVC) filter:**
  - Placement of IVC filters is reserved for high-risk patients with contraindications to anticoagulant therapy.
  - An IVC filter is not a substitute for prophylaxis.
  - Indications for mechanoprophylaxis (IVC filter):
    - Indicated for PE prophylaxis. Does not prevent DVT formation.
    - Failed anticoagulant prophylaxis
    - Contraindication to chemoprophylaxis
    - High-level complete tetraplegia with poor cardiopulmonary reserve
- **Duration of DVT prophylaxis in SCI:**
  - Incomplete SCI and ambulating:
    - Can continue until discharge (but can be continued if soon after injury or other medical comorbidities are present)
  - Complete SCI:
    - Continue at least 8 weeks postinjury in uncomplicated complete motor injury.
    - Continue 12 weeks postinjury, or can consider until discharge from rehabilitation (if >12 weeks) in persons with complete motor injury and other risk factors (i.e., lower limb fracture, history of thrombosis, cancer, heart failure, obesity, or age >70).
- **DVT management:**
  - Therapeutic anticoagulation is similar to the non-SCI population.
  - If documented proximal DVT, treatment continued for 6 months
  - If there is a contraindication to anticoagulant therapy or significant risk of hemorrhage, IVC filter is used to prevent PE.

Functional Electrical Stimulation (FES) in SCI Has Two General Uses:

- As exercise to avoid complications of muscle inactivity
- As a means of producing extremity motion for functional activities:
  - FES can be used to provide a cardiovascular conditioning program
  - Increase muscle bulk, strength, and endurance
  - Produce motion for UE activity, bladder function, standing, and ambulation

SCI, spinal cord injury; UE, upper extremity.

PAIN IN THE SPINAL CORD INJURY PATIENT

- Prevalence in SCI: 60% to 80%; 18% to 44% functionally disabling
- Many are willing to trade pain relief for loss of bladder, bowel, or sexual function.
- There are no consistent associations between the presence of pain and SCI characteristics such as sex, level of injury (paraplegia vs. tetraplegia), and completeness of injury (complete vs. incomplete).
- The International Spinal Cord Injury Pain (ISCIP) classification is recommended and organizes the different pain types (nociceptive and neuropathic) seen after SCI (see **Table 7–10**) and has been incorporated into the International SCI Pain Basic Data Set (ISCIPBDS):
  - Nociceptive pain: From bone, ligaments, muscle, skin, other organs
  - Neuropathic pain: From peripheral or central neural tissue damage

TABLE 7–10 The International Spinal Cord Injury Pain (ISCIP) Classification

TIER 1: PAIN TYPE	TIER 2: PAIN SUBTYPE	TIER 3: PRIMARY PAIN SOURCE AND/OR PATHOLOGY EXAMPLES
Nociceptive pain	Musculoskeletal pain	Lateral epicondylitis, comminuted femur fracture, muscle spasm
	Visceral pain	Abdominal pain owing to bowel impaction, cholecystitis
	Other nociceptive pain	Migraine headache, surgical skin incision
Neuropathic pain	At-level SCI pain	Spinal cord compression, nerve root compression
	Below-level SCI pain	Spinal cord ischemia, spinal cord compression
	Other neuropathic pain	Carpal tunnel syndrome
Other pain	-	Fibromyalgia, complex regional pain syndrome type I, interstitial cystitis, irritable bowel syndrome
Unknown pain	-	

SCI, spinal cord injury.

NOCICEPTIVE PAIN (MUSCULOSKELETAL/VISCERAL)

- More common than neuropathic pain
- Damage to organs (i.e., nonneural tissue) including bone, ligaments, muscle, skin, other viscera

Musculoskeletal Nociceptive Pain

- Approximately 70% of individuals with chronic SCI report pain in their UEs, notably in the shoulder. Additionally, back pain is a common complication.
- 🕮 The shoulder is the most commonly affected joint, usually due to use as a weight-bearing joint and overuse syndromes:
  - Tendonitis/bursitis
  - Rotator cuff impingement/tear
  - Impingement syndrome
  - Subacromial bursitis
  - Capsulitis
  - Myofascial pain
- **Risk factors:**
  - Tetraplegia > paraplegia
  - ↑ with time from injury, older age
  - Associated with mobility devices including manual WC use, ambulation aids, and transfers. Causes include overuse, overhead activities, inflexibility, and muscle imbalances.
  - Shoulder pain is a symptom, NOT a diagnosis!
- **Etiology:**
  - Intrinsic versus extrinsic sources
  - C-spine changes
  - HO
  - Abdominal pathology (with lesions above T7)
  - Cervical root compression
  - Syringomyelia
- **Diagnosis:**
  - Complete history and physical exam, including functional assessment, ROM, flexibility, and sensation testing
  - Radiologic and electrodiagnostic testing as needed

- **Treatment:**
  - Rest to ↓ acute pain
  - Medications (NSAIDs, etc.)
  - Physical therapy with education of proper technique for all activities
  - Modalities
  - Injections as needed
  - Address secondary disabilities
- **Therapy:**
  - PT for the shoulder should focus on stretching anterior and strengthening posterior musculature:
    - **Stretch** anterior shoulder, adductors, and external rotators (often become hypertrophied and contracted).
    - **Strengthen** posterior shoulder girdle muscles including rotator cuff as well as shoulder stabilizers ([trapezius, rhomboids, levator scapulae, serratus anterior] and adductors) to maintain a balanced shoulder.
  - Proper posture and WC setup
  - Correct and improve ADLs. Avoid activities that promote shoulder impingement.
  - Train in proper transfer and weight shift activities.

Visceral Nociceptive Pain

- Pain generated from visceral structures located in the thorax or abdomen
- Dull, aching, or cramping type of pain is more characteristic of visceral pathology or dysfunction (e.g., infection or obstruction).
- Should always consider the differential diagnosis:
  - Examples: UTI, ureteral calculi, impaction, appendicitis

NEUROPATHIC PAIN IN SPINAL CORD INJURY

- Develops in a significant portion of the SCI population
- Damage to peripheral or central neural tissue
- Characterized as "burning," "tingling," "shock-like," "cold," and so on
- Approximately one-third have severe disabling pain.
- No correlation with injury level or severity
- Correlates with increasing age at injury:
  - Peaks in 30 to 39 age group and then again at age >50:
- A thorough evaluation should take place to search for potentially treatable causes of neuropathic pain, such as nerve root or spinal cord compression, tethering, or syringomyelia. This is especially important if the pain first appears >1 year after injury.
- **Treatment:**
  - Treatment interventions for neuropathic pain can be divided into oral and topical medications, procedural interventions, surgical interventions, PT, including exercise, passive and stimulation therapies, as well as relaxation and psychotherapy.
  - Identify if underlying cause of pain
  - Evaluate psychosocial factors
  - Evaluate functional status (activity levels)
  - Interdisciplinary treatment recommended
  - **Medications:**
    - Oral medications are mainly of two groups: Anticonvulsants (that work via suppression of aberrant electrical activity) and antidepressants (via serotonin and NE).
    - These may cause CNS-related side effects (e.g., dizziness, somnolence, ataxia) and it is important to find a balance between side effects and pain relief. Guidelines for pharmacologic management include the CanPain SCI Clinical Practice Guidelines (**Table 7–11**).

TABLE 7–11 The CanPain SCI Clinical Practice Guidelines for Rehabilitation Management of Neuropathic Pain After Spinal Cord Injury

RECOMMENDATION	FIRST LINE	SECOND LINE	THIRD LINE	FOURTH LINE
Strong recommendation	Pregabalin Gabapentin Amitriptyline (TCAs)	Oxcarbazepine Lamotrigine (for incomplete cord lesion) Tramadol		
Weak recommendation			tDCS combined tDCS + VI Botulinum toxin A	TENS Oxycodone DREZ ablation Cannabinoids

DREZ, dorsal root entry zone; SCI, spinal cord injury; TCAs, tricyclic antidepressants; tDCS, transcutaneous direct current stimulation; TENS, transcutaneous electrical nerve stimulation; VI, visual illusion.
Source: From Loh E, Mirkowski M, Agudelo AR, et al. The CanPain SCI clinical practice guidelines for rehabilitation management of neuropathic pain after spinal cord injury: 2021 update. *Spinal Cord* 2022;60:548–566. doi:10.1038/s41393-021-00744-z.

- Pregabalin is the only medication approved for the indication of neuropathic pain in SCI.
- Topical capsaicin, lidocaine, diclofenac
- Other: N-methyl-D-aspartate (NMDA) antagonists, clonidine, tramadol/opioids
- Nonpharmacologic interventions include acupuncture, transcutaneous electrical nerve stimulation (TENS), massage, hypnosis, and biofeedback; visual imagery may be helpful. Biopsychosocial approach including cognitive behavioral therapy may also help with coping skills.

Upper Extremity Compression Neuropathies

- Nearly two-thirds of patients with SCI develop UE compression neuropathies.
- Increases with the length of time from injury and includes most commonly median and ulnar neuropathies
- 🕮 The incidence of carpal tunnel syndrome (CTS) is between 21% and 65% in persons with paraplegia, who are more affected than those with tetraplegia:
  - CTS is due to recurrent stress from transfers, WC propulsion, and pressure relief.
  - Treatment includes analgesics, NSAIDs, splinting (especially at night), injections (anesthetic and/or corticosteroid), physical modalities (US, friction massage, etc.), and education regarding transfers to avoid end range stress.
  - Padded glove use may decrease the trauma of WC propulsion.
  - Surgical release may be required, with the postoperative recovery time being weighed against the long-term benefits of the procedure.
- Ulnar neuropathy is also common.
- 25% bilateral UEs are affected.

Posttraumatic Syringomyelia: Posttraumatic Cystic Myelopathy

- The most common cause of progressive myelopathy after SCI is posttraumatic syringomyelia.
- The pathogenesis of posttraumatic syringomyelia is not entirely understood.
- The most widely accepted mechanism includes subarachnoid scarring with resultant obstruction of CSF flow. Reactive ependymal cell proliferation causes segmental closures in the canal, with distention and movement of CSF into the gray matter of the spinal cord. Arachnoid adhesions affect the movement of CSF, causing progression of the syrinx.
- Other theories suggest that ischemia in the watershed regions of the spinal cord causes cell death and extracellular fluid to coalesce into a syrinx.
- Findings: Cavitation of the spinal cord usually begins at the level of the injury, specifically in the gray matter between the dorsal horns and posterior columns. Cavity formation may be secondary

to liquefaction of the spinal cord or from central hematoma present at the initial injury. Other initiating factors include spinal kyphotic deformity, residual canal stenosis, or compression.

- 🕮 The lesion progresses in a cephalad and caudal direction. As the lesion progresses and compromises more nerve fibers, symptoms may become more apparent.
- Presents clinically in up to 8% of patients but is more frequently first seen on MRI as an elongated cavity in a much higher percentage of cases.
- Posttraumatic syringomyelia may develop at any time, from 2 months to decades postinjury.
- 🕮 Most common presenting symptom is pain:
  - Aching or burning, often worse with coughing, sneezing, straining, and usually in the sitting rather than in the supine position
- 🕮 The earliest sign is an ascending loss of deep tendon reflexes.
- Ascending sensory level loss is also common:
  - Dissociated sensory loss (impaired pain and temperature sensation but intact touch, etc.)
- Weakness occurs but rarely in isolation.
- Additional findings: Spasticity and/or hyper- or hyporeflexia, hyperhidrosis, AD, loss of reflex bladder, worsening HO, new Horner's syndrome, reduced respiratory drive, diaphragmatic paralysis, cranial nerve dysfunction
- 🕮 Diagnosis: MRI with gadolinium is the gold standard.
- Close monitoring (neurologic examinations and MRI) with symptomatic relief is important.
- Activity restrictions:
  - Avoid maneuvers that ↑ intrathoracic/abdominal pressure, such as weightlifting; anterior weight shifts; and Valsalva, Crede, and quad coughing, especially if these exacerbate symptoms.
- Rehabilitation interventions as needed (i.e., functional training and adaptive equipment)
- Treat with pain medications.
- Surgical intervention:
  - Indications for surgery include progressive neurologic decline or severe intractable pain.
  - The most common surgical intervention is shunting (syringo-subarachnoid, syringo-pleural, or syringo-peritoneal) followed by reconstruction of the subarachnoid space with dissection of arachnoiditis/meningeal scarring, and duraplasty.
  - Yields improved strength and pain control in most, but sensory recovery is not usually as favorable.
  - Recurrence of symptoms is common (approximately 50%).

Charcot Joints and Spine

- **Charcot joints:** A destructive arthropathy of joints due to impaired pain perception or position sense
  - Classically been associated with the tabes dorsalis complication of tertiary syphilis (most commonly associated now with diabetes) but also described in SCI
  - Etiology suspected to be a loss of sensation of deep pain or of proprioception that affects the joints' normal protective reflexes, allowing trauma (especially repeated minor episodes) and small periarticular fractures to pass unrecognized
- **Charcot spine:** Spinal trauma and analgesia below the level of injury make SCI patients particularly prone to insensate joint destruction. This can progress to the point of spinal instability, neurologic loss of function, and can become a significant source of pain and deformity in the patient with chronic SCI.

SURGICAL INTERVENTIONS OF THE UPPER EXTREMITY IN TETRAPLEGIA

- Surgical interventions can be used in conjunction with traditional therapies to improve functional mobility in appropriately selected persons with tetraplegia.
- A discussion of anticipated outcomes and the postoperative recovery plan prior to procedures allows for establishment of realistic expectations. Studies suggest that benefits of transfer procedures are maintained and patient satisfaction remains high.

- Procedures (alone or more commonly in combination):
  - Tendon transfers
  - Arthrodesis of interphalangeal (IP) joints of the thumb
  - Implantation of UE neuroprostheses
  - Nerve grafting
- Goal: To improve motor function.
- A **tendon transfer procedure** is when a functioning muscle and its tendon are detached from its normal insertion and rerouted into a different muscle that helps perform the function that needs restoration. In this way, when the donor muscle contracts, it will now power a new and desired motion.
- In a **nerve transfer procedure**, a motor fascicle/branch from a functioning and voluntarily controlled muscle, typically from a major peripheral nerve, is cut and coapted into the motor fascicle/branch of a paralyzed muscle. As with tendon transfers, the original function donor nerve (or muscle) is lost for the sake of a newer and more desired function, so the function of the donor must either be functionally unimportant or duplicative.

Tendon Transfers: Timing and Preop Evaluation

- Timing: Tendon transfers are generally delayed for 1 year to allow neurologic stabilization.
- Components of preoperative evaluation for tendon transfers:
  - Strength: Including shoulder (needed for hand placement). See Modified International Classification of the Upper Limb in Tetraplegia given in Table 7–12.
  - Sensation: Weber two-point discrimination test at pulp of thumb (grade of O-Cu or simply O) given based on ability or inability to distinguish objects <10 mm apart,

TABLE 7–12 Modified International Classification of the Upper Limb in Tetraplegia (Regarding Tendon Transfer Surgery)

GROUP	FUNCTIONAL MUSCLES (GRADE 4 OR 5)
Motor	
0	Weak or absent BR (grade 3 or less)
1	BR
2	BR, ECRL
3	BR, ECRL, ECRB
4	BR, ECRL, ECRB, PT
5	BR, ECRL, ECRB, PT, FCR
6	BR, ECRL, ECRB, PT, FCR, finger extensors
7	BR, ECRL, ECRB, PT, FCR, finger extensors, thumb extensors
8	BR, ECRL, ECRB, PT, FCR, finger extensors, thumb extensors, finger flexors
9	Lacks intrinsics only
Sensory	
O:	Two-point discrimination in thumb >10 mm
Cu:	Two-point discrimination in thumb <10 mm

BR, brachioradialis; ECRB, extensor carpi radialis brevis; ECRL, extensor carpi radialis longus; FCR, flexor carpi radialis; PT, pronator teres; O, inability to distinguish objects <10 mm apart; Cu, ability to distinguish objects <10 mm apart.

respectively. A grade of O-Cu is predictive of sufficient proprioception to allow hand function without visual cues for ease of ADLs.
  - ROM: Including shoulder (needed for hand placement, and in consideration for posterior deltoid or biceps to triceps transfers); hand/wrist (ROM should be full for all procedures involving the hand)
  - Spasticity: Should be minimal or absent in proposed transfer muscles
  - Behavioral evaluation: Extremely important, as recovery phase involves immobilization, restriction of previously noted activities, and potential need for more assistance from caregivers
- Classification is utilized to standardize communication regarding tendon transfer surgery.
- Functional muscle strength is considered grade 4 or 5 (unlike the International Standards exam, where motor level is based on "normal" strength of 3 or greater) because muscles being transferred generally lose one strength grade.
  - As a result, muscles with a grade of 3 or less are not considered for transfer, as they would likely not have sufficient strength for functional tasks following the surgery.

🕮 Tendon Transfer Procedures by Spinal Level

- **C5 level:**
  - *Brachioradialis to extensor carpi radialis brevis (ECRB)*: Restore wrist extension (improve lifting objects, feeding, grooming, hygiene).
  - *Deltoid to triceps*: Provide elbow extension (stabilize sitting and transferring, reach overhead against gravity, grooming, hygiene, pressure relief, writing speed and clarity, feeding).
- **C6 level:**
  - *Moberg "key grip" procedure*: Restore lateral or "key" grip (improve grooming, eating, writing, desktop skills).
  - *Brachioradialis (or other active muscles) to flexor pollicis longus (FPL [or finger flexors])*. Restore lateral pinch (FPL) or grasp (finger flexors). Provides better function and preferable to Moberg procedure.
  - *Posterior deltoid to triceps*: Recommended prior to hand reconstruction, or simultaneous without diminishing outcomes. *Biceps to triceps* tendon transfers are also performed.
  - *Rerouting of biceps brachii around radial neck*: Correct supination contracture of forearm that may occur in C5 and C6 motor injuries.
- **C7 level:**
  - *Brachioradialis to FPL*: Restore thumb flexion.
  - *Extensor carpi radialis longus (ECRL) or flexor carpi ulnaris (FCU) to flexor digitorum profundus (FDP)*: Restore finger flexion.
- **C8 level:**
  - Intrinsic minus or "claw hand" may be addressed with lumbrical bar, preventing hyperextension of metacarpophalangeals (MCPs) to improve function.
  - Surgery rarely indicated
- **Postoperative management:**
  - Casted extremity usually immobilized for a few weeks and then changed to removable splints. The time of immobilization depends upon the specific technique and surgeon preferences.
  - Edema controlled with elevation techniques
  - Removable thermoplastic splint fabricated to protect transfer site from overstretching once cleared for ROM
  - Scar management with scar mobilization and desensitization techniques
  - Biofeedback and electrical stimulation for muscle re-education
  - Activities creating heavy resistance to tendons eliminated until after 3 months

Nerve Grafting

- Unlike tendon transfers, where there is noted improvement a few months postsurgery, progress with nerve grafting may not be seen for at least 1 year. However, the immobilization period postoperatively is much shorter, and ultimate strength and function may be improved:

- Numerous techniques for the UEs have been described and are beyond the scope of this text (Fox et al., 2025).

SPASTICITY

- Spasticity presents as an abnormality of muscle tone and is common in SCI individuals. It becomes clinically apparent as spinal shock resolves.
- See Chapter 12, Associated Topics in Physical Medicine and Rehabilitation, section on Spasticity for further detail.

DUAL DIAGNOSES: TRAUMATIC BRAIN INJURY WITH SPINAL CORD INJURY

- 🕮 The incidence of concomitant TBI in those with a primary SCI is reported between 24% and 74%.
- Historical factors, such as mechanism of injury (e.g., MVC), loss of consciousness, being male, higher NLI, presence of posttraumatic amnesia (PTA), and impaired initial Glasgow Coma Scale (GCS) score, are risk factors for concomitant TBI.
- Deficits may be seen in attention, concentration, and memory, and can interfere with new learning and problem-solving.
- Medical management of problems common to the SCI population, such as pain, DVT prophylaxis, spasticity, and NLUTD, requires special consideration in the dual diagnosis patient.
- Special care should be taken to utilize medications with minimal cognitive impact. Medications routinely used in SCI may have an impact on the recovering brain (i.e., baclofen and benzodiazepines).
- Elevated BP may signal the presence of AD in those with SCI at or above the T6 level. However, those with TBI may experience transient elevation of BP secondary to a centrally driven "sympathetic storm."

PSYCHOLOGICAL ISSUES IN THE SPINAL CORD INJURY PATIENT

- 🕮 Depression occurs in 20% to 45% of those injured and usually occurs within the first month.
- Although not all individuals with SCI will experience depression, depressive disorders are the most common form of psychological distress following SCI.
- Individuals with depression have less functional independence at discharge, experience more pressure injuries, have increased pain and more general health problems, and engage in less participation in the community.
- 🕮 The suicide rate for individuals with SCI is approximately 3 to 5× the age- and sex-specific suicide rate in the United States:
  - 🕮 It is the leading cause of death in individuals with SCI in the youngest age groups.
  - The greatest risk is within the first 5 years postinjury.
  - Risk factors include previous suicide attempts, preinjury depression, individual coping styles, poor self-esteem, younger age, loss of male identity, feelings of guilt or shame, alcohol use, active involvement in causation of injury, and undiagnosed mild TBI.
  - Although individuals with paraplegia and more incomplete injuries appear to have a higher rate of suicide, level of injury is undetermined as a risk factor.
- Treatment includes pharmacology and counseling. The most commonly prescribed medications for depression in SCI are selective serotonin reuptake inhibitors (SSRIs). A psychiatrist should be consulted if the patient has suicidal ideation or presents with psychotic features, and/or the patient does not respond to one or two of the trials of antidepressant medications.
- Anxiety and posttraumatic stress disorder (PTSD) have been reported in up to 20% of persons with SCI. Symptoms of PTSD may be higher in those with features of depression and/or anxiety.
- Substance abuse is frequently encountered in SCI. Alcohol and drug misuse contributes to SCI by increasing risk taking while intoxicated; hampers learning and rehabilitation gains; interferes with self-care; places persons at risk for complications; contributes to depression, mortality, and morbidity; limits long-term outcomes; and impacts capacity for independent living.

PRESSURE INJURIES

- Approximately 25% of acute SCI patients develop a pressure injury, with up to 95% of patients developing one at some time during their life.
- Skin issues are the second most common etiology for rehospitalization in chronic SCI.
- Pressure injuries are classified (according to the extent of tissue damage) utilizing the National Pressure Injury Advisory Panel (NPIAP) staging system, with the latest new revisions from 2019.

NATIONAL PRESSURE INJURY ADVISORY PANEL STAGING OF PRESSURE INJURIES (TABLE 7–13)

- 🕮 **Location (Figure 7–31):**
  - Pressure injuries most commonly develop over bony prominences.
  - 🕮 The most common location in persons with SCI within the first 2 years is the sacrum, followed by the ischium, heels, and trochanters.
  - After 2 years, the ischial tuberosities are the most common site of development.
  - 🕮 In children, the occiput is the most frequent site.
- **Risk factors:**
  - 🕮 The most important risk factors for developing pressure injuries are persistent **pressure** and **shear** forces.
  - Others include higher level and greater severity of the injury (complete tetraplegia the highest, followed by complete paraplegia, then incomplete tetraplegia and paraplegia, respectively), immobility, unemployment status, lower educational achievement, tobacco and alcohol use, history of a prior pressure injury, poor nutritional status, anemia, incontinence, smoking, and possibly psychosocial issues (e.g., depression):
    - The Braden Scale is the most common tool used by clinicians to assess risk for pressure injury development.

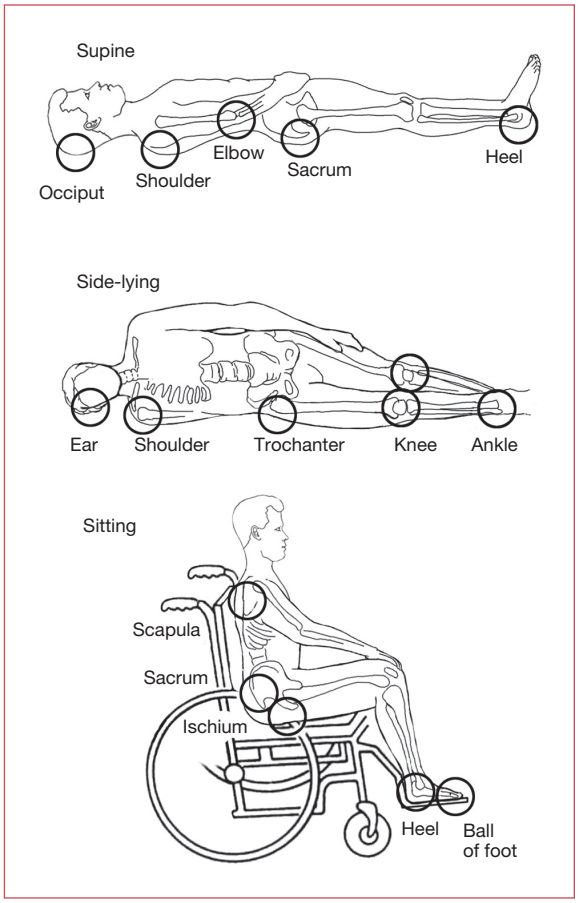
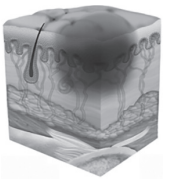
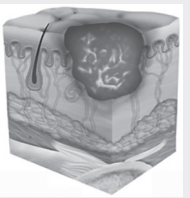
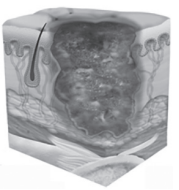
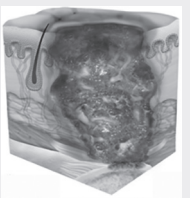
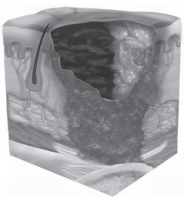


FIGURE 7–31 Common locations of pressure injuries.

🕮 MECHANISMS OF DEVELOPING A PRESSURE INJURY

- 🕮 Local soft tissue ischemia results due to prolonged pressure over bony prominences that exceed supracapillary pressure:
  - **Ischemia:** Lack of adequate blood supply to the tissue:
    - Frequently associated with hyperemia in the surrounding tissue
    - Increased local O_2 consumption occurs

TABLE 7–13 NPIAP Staging of Pressure Injuries

STAGE	DESCRIPTION	
Stage 1	Intact skin with localized area of nonblanchable erythema, which may appear differently in darkly pigmented skin. Blanchable erythema or changes in sensation, temperature, or firmness may precede visual changes. Color changes DO NOT include purple or maroon discoloration; these may indicate deep tissue pressure injury.	Stage 1 Pressure injury - lightly pigmented 
Stage 2	Partial-thickness loss of skin with exposed dermis. The wound bed is viable, pink or red, moist, and may also present as an intact or ruptured serum-filled blister. Adipose (fat) and deeper tissues are not visible. Granulation tissue, slough, and eschar are not present. This stage should not be used to describe moisture associated with skin damage or dermatitis, medical adhesive-related skin injury, or traumatic wounds (e.g., skin tears, burns, abrasion).	Stage 2 Pressure injury 
Stage 3	Full-thickness loss of skin, in which adipose is visible in the ulcer and granulation tissue and epibole (rolled wound edges) are often present. Slough and/or eschar may be visible. The depth of tissue damage varies by anatomic location; areas of significant adiposity can develop deep wounds. Undermining and tunneling may occur. Fascia, muscle, tendon, ligament, cartilage, and/or bone are NOT exposed. If slough of eschar obscures the extent of tissue loss, this is an Unstageable Pressure Injury.	Stage 3 Pressure injury 
Stage 4	Full-thickness skin and tissue loss with exposed or directly palpable fascia, muscle, tendon, ligament, cartilage, or bone in the ulcer. Slough and/or eschar may be visible. Epibole (rolled edges), undermining, and/or tunneling often occur. Depth varies by anatomic location. If slough or eschar obscures the extent of tissue loss, this is an Unstageable Pressure Injury.	Stage 4 Pressure injury 
Unstageable	Full-thickness skin and tissue loss in which the extent of tissue damage within the ulcer is obscured by slough or eschar. If slough is removed, a Stage 3 or Stage 4 pressure injury will be revealed. Stable eschar (i.e., dry, adherent, intact without erythema or fluctuance) on the heel or ischemic limb should not be softened or removed.	Unstageable 

(*continued*)

TABLE 7–13 NPIAP Staging of Pressure Injuries (*continued*)

STAGE	DESCRIPTION	
Deep tissue injury (DTI)	Intact or nonintact skin with localized area of persistent nonblanchable deep red, maroon, purple discoloration or epidermal separation revealing a dark wound bed or blood-filled blister. Pain and temperature change often precede skin color changes. Discoloration may appear differently in darkly pigmented skin. The wound may evolve rapidly to reveal the actual extent of tissue injury, or may resolve without tissue loss. If necrotic tissue, subcutaneous tissue, granulation tissue, fascia, muscle or other underlying structures are visible, this indicates a full-thickness pressure injury (Unstageable, Stage 3, or Stage 4). Do not use deep tissue pressure injury to describe vascular, traumatic, neuropathic, or dermatologic conditions.	Deep tissue pressure injury
Additional Pressure Injury Definitions		
Medical Device-Related Pressure Injury	Medical device-related pressure injuries result from the use of devices designed and applied for diagnostic or therapeutic purposes. The resultant pressure injury generally conforms to the pattern or shape of the device. The injury should be staged using the staging system.	
Mucosal Membrane Pressure Injury	Mucosal membrane pressure injury is found on mucous membranes with a history of a medical device in use at the location of the injury. Due to the anatomy of the tissue, these ulcers cannot be staged.	

NPIAP, National Pressure Injury Advisory Panel.
Source: Adapted from National Pressure Injury Advisory Panel. *NPIAP Pressure Injury Stages*. Updated 2019.

- **Pressure:**
  - 🕮 Prolonged pressure over bony prominences, exceeding supracapillary pressure continuously for 2 or more hours, results in occlusion of the microvessels of the dermis with subsequent tissue ischemia
  - Occlusion of microvessels occurs when the force exerted on the vessel wall is greater than the intra-arterial pressure.
  - This results in immediate epidermal ischemia.
  - Ischemia causes hyperemia of the surrounding tissue.
  - 🕮 Muscle is more susceptible to pressure ischemia than skin.
- **Friction (shearing forces):**
  - Removes stratum corneum of the skin
  - Friction mechanically separates the epidermis immediately above the basal cells.
  - Friction is a factor in the pathogenesis since it applies mechanical forces to the epidermis.

PREVENTION OF PRESSURE INJURIES

- Minimize extrinsic factors: Pressure, maceration, and friction.
- 🕮 Decrease duration of pressure forces → the patient should initially be turned and positioned every 2 hours.
- 🕮 Pressure relief (weight shifting) and repositioning should be done for at least 2 minutes at a time, every 15 to 30 minutes when sitting.
- Proper mattress/bed overlay
- Proper cushioning and WC seating

- Pressure mapping can be helpful in establishing an appropriate seating system for the patient to assure that appropriate pressure distribution occurs while sitting and that appropriate weight relief is produced during the pressure relief.

TREATMENT OF PRESSURE INJURIES

- Prevention of pressure injuries should always be the first line of defense.
- Once a lesion has developed, however, treatment should be prescribed to reduce its progression. Extrinsic factors that contributed to the initial formation of the pressure injury should be identified and treated.
- In general, healing will be promoted if the wound remains clean and moist, with debridement of necrotic and infected tissue as needed.
- Stage 1 and 2 pressure injuries are usually nonoperative.
- Stage 3 injuries may require surgical intervention (debridement, muscle/skin flaps).
- Stage 4 pressure injuries may require surgery.
- Medications/vitamin supplementation can be used, particularly in severe cases (European Pressure Ulcer Advisory Panel, National Pressure Injury Advisory Panel and Pan Pacific Pressure Injury Alliance, 2019).
- A bone biopsy is the gold standard for the diagnosis of a bacterial infection and is the only method that should be used to direct antibiotic therapy.
- Although rare in SCI patients, carcinomas arising within a chronic pressure injury, termed Marjolin ulcers, are highly aggressive and frequently follow an unremitting and usually fatal course. A sudden change in a long-standing indolent pressure injury (average latency of 20 years) should raise suspicion.

Wound Healing

- To heal, the wound must be clean and free of necrotic tissue:
  - Mechanical, enzymatic, or surgical debridement (sharp debridement)
- Cleansing solutions:
  - Isotonic saline for most wounds
  - The stronger the cleanser, the more toxic.
  - As the wound becomes cleaner, the strength of the cleansing solution should be decreased.
  - Technique: No hard rubbing or scrubbing
- Whirlpool therapy
- Pulsatile irrigation (<15 psi) dressings: Multiple types, depending on if wound is dry, moist, or necrotic:
  - To decrease wound maceration, apply petroleum-type gauze to the wound edges or a stomal adhesive wafer around the wound.
  - To prevent abscess formation, cavities should be loosely packed.
  - For wounds with minimal to mild exudates, dressing choices can include a hydrocolloid, polyurethane foam, or saline gauze dressing.
  - With moderate-heavy exudates, a calcium alginate dressing is suggested.

Wound Care Modalities

- Based on the clinical evidence available, the Agency for Health Care Policy and Research (AHCPR) found only electrical stimulation merited recommendation. Used for nonhealing pressure injuries:
  - ↑ circulation, granulation, with ↓ bacterial count
  - Not used if cellulitis is present; pacemaker; or metal ions
- The therapeutic efficacy of hyperbaric oxygen; infrared, ultraviolet, and low-energy laser irradiation; and ultrasonography has not been sufficiently established for recommendation.
- Wound vacuum-assisted therapy system—an open-cell foam into the wound, sealing the site and applying subatmospheric pressure (125 mmHg below ambient):

  - ↑ blood flow into the wound and adjacent tissue leads to ↑ O_2 and nutrient delivery and ↑ clearance of bacteria from infected wounds, resulting in an environment that promotes wound healing

Surgical Interventions

- Musculocutaneous flap: Most common and most often recommended in severe pressure injuries in SCI. In this type of surgery, the skin is transferred with underlying muscle/blood vessels.
- Other types of flaps are being used in SCI and include:
  - Fasciocutaneous flaps (includes just the skin, subcutaneous tissue, and deep fascia)
  - Pedicled perforator flaps (blood supply to the skin and subcutaneous tissue)
- Osteomyelitic bone can be removed.
- Total ischectomy is not recommended as a means of reducing recurrence.
- Rotation flap:
  - Semicircular flap rotating about a pivot point to close a triangular defect
- Transposition flap:
  - Rectangular flap rotates about its base to fill an adjacent defect
- Advancement flap:
  - Moved into a defect without lateral or rotational movement

Postop Management for Musculocutaneous Flap Procedures

- Strict bedrest—at least 3 to 4 weeks
- Vigilant pressure relief and avoidance of shear forces
- Air-fluidized bed:
  - Small ceramic beads in motion simulate the movement of fluid.
  - Temperature adjustable, good pressure relief, absorption of wound fluids away, bacteriostatic capabilities of the beads
- Sitting time: If no problem after immobilization:
  - Slowly increased: Start 15 minutes daily and increase by 15 minutes two times a day (BID).
  - Monitor flap closely afterward.

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PHYSICAL MODALITIES, THERAPEUTIC EXERCISE, EXTENDED BEDREST, AND AGING EFFECTS

8

Thomas E. Strax, MD • Priscila Gonzalez, MD • Steven V. Escaldi, DO • Sara J. Cuccurullo, MD • James Yun, MD

PHYSICAL MODALITIES

Physical modalities are used as adjuncts in a comprehensive therapy program that includes exercise, medication, adaptive equipment, and patient education. They use physical energy for their therapeutic effects:

- Thermotherapy (heat and cold modalities)
- Hydrotherapy
- Light therapy (ultraviolet [UV] radiation, laser)
- Electrotherapy
- Manual therapy (traction, therapeutic massage, manipulation, mobilization)
- Pressure
- Acupuncture

THERMOTHERAPY

- The amount of energy a tissue gains or loses depends on several factors:
  - Nature of the tissue
  - Agent used
  - Duration of exposure
- Temperature has an effect on:
  - **Viscosity—**blood viscosity decreases as temperature increases (Buono et al., 2016).
  - **Nerve signal conduction**—heat increases nerve conduction velocity; cold decreases it.
  - **Blood flow**—heat increases arterial and capillary blood flow; cold decreases blood flow.
  - **Collagen extensibility**—heat increases tendon extensibility and collagenase activity; cold decreases enzyme activity.
  - 🕮 **Combined use of heat and stretching** has been shown to significantly increase tendon extensibility (Weber & Hoppe, 2011).
- Temperatures >113°F to 122°F (45°C–50°C) or <32°F (0°C) can injure tissue.

A. Heat

Therapeutic uses for heat are based on:

- Hyperemia
- Analgesia
- Hyperthermia

- Decreased muscle tone
- Increase in collagen elasticity

Applications for heat therapy:
- Generally used for subacute and chronic processes
- Reduction of muscle spasms
- Pain reduction (myofascial, low back, neck, postherpetic neuralgia)
- Reduction in joint stiffness and contractures
- Arthritis and collagen vascular diseases
- Chronic inflammation
- Superficial thrombophlebitis

Contraindications to heat therapy:
- 🕮 Ischemia, that is, arterial insufficiency:
  - Metabolic requirement of the limbs is increased with the use of heat.
  - Note: For every 10°F increase in skin temperature, there is a 100% increase in metabolic demand.
- Bleeding disorders (e.g., hemophilia) or hemorrhage—there is an increased arterial and capillary blood flow with heat.
- Impaired sensation—central (e.g., spinal cord injury, stroke) or peripheral neuropathies may predispose individuals to burns.
- Inability to communicate or respond to pain—for example, dementia and aphasia.
- Malignancy—may increase tumor growth and spread.
- Acute trauma or inflammation—diffusion across membranes is increased.
- 🕮 Scar tissue—elevation of temperature increases the tissue's metabolic demand. Scar tissue has inadequate vascular supply and is unable to provide an adequate vascular response when heated, which can lead to ischemic necrosis.
- Edema—diffusion across membranes is increased.
- Atrophic skin
- Poor thermal regulation

Mechanisms for heat transfer:
- Heat modalities utilize three main methods of heat transfer:
  1. Conduction
  2. Convection
  3. Conversion
- Conduction and convection heat transfer modalities focus on superficial heating, whereas deep heating modalities utilize conversion to achieve greater tissue penetration.

Conduction

Conduction is the transfer of heat between two bodies at different temperatures through direct contact. Heat transfer occurs without movement of the conducting body.
- Examples:
  - Hot water
  - Paraffin bath
  - Hot packs (hydrocollator packs)
  - Kenny packs
  - Heating pads

Convection

Convection is the transfer of heat that occurs by circulation (typically liquid or gas) over the surface of a body. Fluid flow increases the temperature gradient between the surfaces, increasing heating and cooling. It is more intense than conduction.
- Examples:
  - Fluidotherapy
  - Contrast baths
  - Hydrotherapy (whirlpool)

Conversion

Conversion is the transfer of heat through conversion of electromagnetic radiation (nonthermal energy) into heat energy.

- Examples:
  - Radiant heat (heat lamps)—superficial heat
  - Ultrasound (US)
  - Shortwave diathermy (SWD)
  - Microwave diathermy

SUPERFICIAL VERSUS DEEP HEAT

Superficial Heat

Superficial heat can achieve tissue heating up to 1 to 2 cm in depth. It can be used to heat joints with little soft tissue covering (hand, foot). It can also cause a deeper effect through reflex mechanisms (e.g., for relief of muscle spasms). Superficial heating modalities achieve heat transfer through conduction, convection, or conversion.

Conduction Agents

- Hot packs:
  - Hydrocollator: Canvas bags filled with silicon dioxide immersed in tanks of heated water (166°F/74.5°C).
  - Applied over several layers of insulating towels usually for 30 minutes at a time.
  - Lehmann et al. (1966): Hot pack applied to posterior thigh increased temperature to 3.3°C (37.94°F) at 1 cm depth and 1.3°C (34.3°F) at 2 cm depth.
  - Advantages: Low cost, minimal maintenance, long life patient acceptance, ease of use.
  - Disadvantages: Prolonged superficial heat can produce temporary or permanent skin mottling—erythema ab igne. This condition is characterized by reticular pattern of erythema, telangiectasia, and hyperpigmentation.
- Kenny packs—wool cloths soaked in 60°C water, then spun dried. These cool rapidly and require repeated applications.
- Heating pads:
  - Available as electric pads and pads with circulating heated fluid, such as water.
  - Peak temperature is 125°F (52°C).
  - The temperature is maintained at a constant level, with no spontaneous cooling.
  - If used with moist towels, there is a potential risk for electrical shock.
  - Potential exists for burns. This is common in patients with decreased adipose tissue.
  - Generally used for periods of 20 minutes.
  - Caution patients about falling asleep on heating pads due to potential for burns.
- Paraffin bath:
  - Paraffin wax and mineral oil in a 7:1 or 6:1 ratio heated to 126°F to 130°F (52.2°C–54.4°C).
  - Commonly used in irregular surfaces, such as distal extremities.
  - Three methods are available for use:
    1. Dipping (most common method): Body part is dipped in a paraffin bath and then quickly removed, allowing the paraffin to cool and harden. Approximately 7 to 12 dips are done, followed by wrapping in wax paper/plastic, which is covered by towels or insulated mitts. Application time is 20 to 30 minutes. A decrease in temperature is noted after 15 to 20 minutes postdip.
    2. Immersion: Serial dips are done, forming a thin shell, which is followed by immersion in the paraffin bath for approximately 30 minutes. This process provides more vigorous heating than dipping.
    3. Brushing: A brush is used to apply paraffin to larger body parts or parts that are difficult to manage with the bath (e.g., ankle).
  - Abramson et al. (1967): 5.5°C temperature increase in forearm subcutaneous tissue and 2.4°C increase in brachioradialis muscle with paraffin use.
  - Patients have good compliance, and home units are available.
  - Uses: Contractures, rheumatoid arthritis (RA), scleroderma.

Convection Agents

- Fluidotherapy:
  - Hot air is blown through a container holding fine cellulose particles (bed of beads or corn husks), which produces a warm air–fluid mixture with properties similar to liquid.
  - Advantages: Massage action of the turbulent solid–gas mixture; freedom to perform range of motion (ROM) activities.
  - Good for hands and feet.
  - Agitation level and temperature can be controlled. The typical temperature range is 115°F to 120°F (46.1°C–48.9°C).
  - Study performed by Borell et al. (1980) measured temperature in different body areas following 20 minutes of fluidotherapy at 118°F (47.8°C), with the following results:
    - Peak temperature of hands and joint capsule is 107.6°F (42°C).
    - Peak temperature of foot and joint capsule is 103.1°F (39.5°C).
  - Fluidotherapy should be avoided on infected wounds, and burn precautions should be maintained.
- Hydrotherapy:
  - External use of water to treat a physical condition. Water can be used to produce convective heating or cooling, massage, and gentle debridement.
  - Unit size, water temperature, agitation intensity, and solvent properties can be adjusted to meet treatment goals.
  - Give treatment for approximately 10 to 20 minutes, depending on the patient's cardiopulmonary tolerance.
  - In general, temperatures from 91.4°F to 96.8°F (33°C–36°C) are considered neutral for wounds and burns and are well-tolerated.
  - Peripheral vascular disease (PVD)—Note: *Increase in skin temperature of 10°C increases metabolic demand by 100%. If PVD is suspected, for example, in a patient with diabetes, ascertain skin temperature before choosing water temperature.*
  - **Whirlpool baths and Hubbard tanks:**
    - Whirlpool baths used for **partial body** immersion
    - Hubbard tanks used for **total body** immersion
    - The water temperature can be selected depending on the amount of body submerged, patient's health, and goals of treatment.
    - Whirlpool temperature for:
      - Upper limbs is 100°F to 105°F (37.8°C–40.6°C)
      - Lower limbs is 100°F to 102°F (37.8°C–38.9°C)
    - Hubbard tank temperatures should be <102.2°F (39°C) to avoid systemic problems (can change core body temperature):
      - Mild heating: 98°F to 98.9°F (36.7°C–37.2°C)
      - Vigorous heating: 100°F to 100.9°F (37.8°C–38.3°C)
    - Contraindications for Hubbard tanks:
      - Bowel or bladder incontinence
      - Skin infections
      - Unstable blood pressure (BP)
      - Uncontrolled epilepsy
      - Acute febrile episodes
      - Upper respiratory infections
      - Tuberculosis
      - Multiple sclerosis
      - Caution in patients with vital capacity (VC) <1 L due to concern over respiratory distress and inability to extract the patient in a timely manner (Weber & Hoppe, 2011)
- Contrast baths:
  - Distal limbs receive alternating heat and cold in a whirlpool tank to produce reflex hyperemia.

- Temperatures range from hot (100.4°F–111°F or 38°C–44°C) to cold (50°F–64.4°F or 10°C–18°C).
- Technique: Begin with warm soaks to the extremity, and then follow with four cycles of alternating 1- to 4-minute cold soaks and 4- to 6-minute warm soaks.
- Uses: RA; complex regional pain syndrome (CRPS), to toughen residual limbs, and for muscular strains and joint sprains.
- Contraindications: Small vessel disease caused by diabetes, arteriosclerotic endarteritis, or Buerger disease.

Conversion Agents

- 🕮 Superficial heat: Radiant heat (infrared lamps):
  - Light energy (nonthermal) is absorbed through the skin and converted to superficial heat.
  - Distance from the lamp to skin is usually 45 to 60 cm (18–24 in.) and is applied for 20 to 30 minutes. Most lamps act as point sources, and their heating effectiveness decreases with the square of their distance from the body ($1/r^2$ law).
  - Used in patients who cannot tolerate the weight of hot packs.
  - Precautions: General heat precautions, light sensitivity (dermal photo-aging), and skin drying; use with photosensitizing medications.

Deep Heat: Diathermy

Tissue temperature can be increased to a depth of 3 to 5 cm or more without overheating subcutaneous tissue or skin. Deep heat transfer is produced by conversion of electromagnetic energy into heat (diathermy) and may penetrate to deep structures such as the ligaments, bones, muscles, and joint capsules. Deep heat modalities include:

1. US
2. SWD
3. Microwave diathermy

1. Ultrasound

Sound waves with frequencies above the audible range (>20,000 Hz) can produce thermal (heating) and nonthermal (cavitation, acoustic streaming, and standing waves) effects.

THERMAL EFFECTS

- US interacts with skin, fat, and muscle during treatment. Heating occurs in all of these tissues as a result of sound wave attenuation and absorption. Its effect is more pronounced at tissue interfaces where sound transmission discontinuities occur.
- US is absorbed and attenuated more in bone, followed by tendon, followed by skin, muscle, and fat.
- Lehmann et al. (1967) found that US produces the highest temperature in cancellous bone (spongy bone).
- 🕮 Absorption (heating) is greatest at the bone–muscle soft tissue interface.
- Thermal effects include increased distensibility of collagen fibers.

NONTHERMAL EFFECTS

- Acoustic cavitation: Gas bubbles produced within the sound field due to turbulence, which by their forced oscillation and bursting are capable of disrupting tissue.
- Acoustic streaming: Unidirectional movement of compressible material or medium due to pressure asymmetries caused by US waves.
- Acoustic streaming and cavitation are associated with wound contraction and protein synthesis.
- Standing waves: In a stationary US field, standing waves produce fixed areas of elevated pressure and rarefaction. They have not been found to have physiologic benefits.

🕮 *ULTRASOUND INDICATIONS*

- Bursitis
- Tendinitis (calcific tendinitis)
- Musculoskeletal pain

- 🕮 Degenerative arthritis and contracture (adhesive capsulitis, shoulder periarthritis, hip contracture). Helps to maintain a prolonged stretch and increases ROM
- 🕮 When addressing the small joints (fingers and toes), it is typically used underwater, but only if water is degassed
- Subacute trauma
- Less established: Scar tissue (keloids), postherpetic neuralgic pain, and plantar warts

🕮 *ULTRASOUND CONTRAINDICATIONS*

- General heat contraindications
- Near the brain, cervical ganglia, spine, and laminectomy sites (can cause spinal cord heating)
- Near the heart and reproductive organs
- 🕮 Near pacemakers—may cause thermal or mechanical injury to the pacemaker
- 🕮 Near tumors
- Gravid or menstruating uterus
- At infection sites
- On contact lenses and eyes (fluid-filled cavity with risk of cavitation and heat damage)
- **Skeletal immaturity**—open epiphysis can be affected with decreased growth due to thermal injury
- Total hip or knee **prostheses with polymethylmethacrylate (PMMA) or high-density polyethylene**. These have a high coefficient of absorption (more than soft tissue), and the prosthesis may loosen due to unstable cavitation in the cement (Lehmann & De Lateur, 1990)
- Arthroplasties—the effect on bony ingrowth arthroplasties is not well-defined, thus the most prudent course is avoiding US over these areas

ULTRASOUND PRECAUTIONS

- Avoid intensities over 3 W/cm^2.
- Use multiple ports over large joints.
- US can be used underwater but only if water is degassed. This can be achieved by putting it into a container and allowing it to sit overnight.

ULTRASOUND PRESCRIPTION

- 🕮 Frequency: 0.8 to 1.1 MHz
- Intensity: 0.5 to 2.0 W/cm^2; maximal intensity is 3.0 W/cm^2, as suggested by the World Health Organization (WHO, 2008); spatial average intensity = power output/effective radiating area
  - 🕮 For tendinitis/bursitis, the average intensity used is 1.2 to 1.8 W/cm^2 (0.5–2.0 W/cm^2 range).
  - Generates temperatures up to 114.8°F (46°C) in deep tissues.
- Duration: 5 to 10 minutes per site; depends on intensity used and size of area being treated
- Heating element must always be in constant motion when applying US to prevent burns.
- US deep heating is superior to microwave and SWD. In general, temperature increases to 113°F (45°C) will be seen at 8 cm depth. Increased temperature will remain for 2 minutes following US application.
- Modes:
  - **Continuous mode:** Thermal effects (heat production)
  - **Pulsed mode:** Mainly nonthermal effects; produces cavitations, media motion, and standing waves
- Techniques (direct and indirect):
  - **Direct (most common):** The applicator is moved slowly over an area of 4 sq. in. in a circular or longitudinal pattern. Uses a coupling media (gel) to accommodate for impedance inconsistencies.
  - **Indirect:** For uneven surfaces (hands and feet). Applicator and body part are immersed in a container of degassed water.

SPECIALIZED USES

- **Phonophoresis:** Technique that utilizes US to drive medications through the skin by increasing cell permeability:
  - Frequency: 1 to 2 MHz
  - Intensity: 1 to 3 W/cm^2 for 5 to 7 minutes
  - Mode: Continuous or pulsed

- Medications: Corticosteroids (1%–10% hydrocortisone and dexamethasone) and/or anesthetics (1% lidocaine)
- Uses: Tendinitis—Achilles, patellar, bicipital; tenosynovitis; epicondylitis (tennis elbow)

2. Shortwave Diathermy

- 🕮 Produces deep heating through the conversion of electromagnetic radio wave energy to thermal energy.
- The Federal Communications Commission (FCC) limits use to 13.56 MHz (22 m wavelength), 27.12 MHz (11 m), and 40.68 MHz (7.5 m).
- 🕮 The most commonly used frequency is 27.12 MHz.
- 🕮 Provides heat over a larger area, as well as deep heat to 4 to 5 cm depth. SWD preferentially heats low impedance tissues, such as the skeletal muscle, blood, and synovial fluid. Therefore, SWD is a good choice if deep muscle heating is the primary goal.
- The heating pattern produced depends on the type of shortwave unit, water content, and electrical properties of the tissue.
- Shortwave units can be *inductive* or *capacitive*.
- ***Inductive coil method:*** Produces high temperatures in water-rich tissues (superficial muscles, skin) via a coiled magnetic field (induction coil). The body acts as a receiver, and eddy currents are induced in the tissues in its field. Applicators are in the form of cables or a drum.
  - Increases tissue temperature 4°C to 6°C above normal.
  - 🕮 Indicated when heat to more superficial muscles or joints with minimal superficial tissue is desired. Muscle tends to become warmer than fatty tissue.
- ***Condensor method:*** Produces high temperatures in water-poor tissues (fat, bone) with low conductivity via rapid oscillation of an electrical field. The treatment area is placed between two capacitor plates to which the shortwave output is applied. The body acts as an insulator in a series circuit in this setup.
  - Indicated for subcutaneous adipose tissue and superficial muscle.
  - More effective in deeper joints (i.e., hip joint).
  - Treatment time is 20 to 30 minutes.
- Precise dosing for SWD is difficult, and pain perception is used to monitor intensity. The best way to monitor frequency depends on the patient's response to warmth.
- Subcutaneous fat temperature rises to 15°C, a 4°C to 6°C increase in muscle at 4 to 5 cm depth (Lehmann et al., 1968).
- Terry cloths are used for spacing and to absorb sweat, which is highly conductive and may cause severe local heating.

SHORTWAVE DIATHERMY INDICATIONS

- Muscle spasms (especially low back)
- Myalgia
- Refractory pelvic inflammatory disease
- Chronic prostatitis

SHORTWAVE DIATHERMY CONTRAINDICATIONS

- General heat precautions
- Metal items (jewelry, pacemakers, metallic **intrauterine devices, surgical implants**), which are excellent electrical conductors and can potentially cause burns; water highly conductive and can have the same effect with resultant severe local heating
- Contact lenses
- Gravid or menstruating uterus
- Skeletal immaturity

3. Microwave Diathermy

- Conversion of microwave electromagnetic energy to thermal energy.
- FCC approved frequencies: 915 MHz (33 cm wavelength) and 2,456 MHz (12 cm).
- Microwaves do not penetrate tissues as deeply as US or SWD.

- Microwave diathermy preferentially heats fluid-filled cavities.
- A lower frequency has a higher depth of penetration and is better for muscle heating.

MICROWAVE DIATHERMY INDICATIONS

- Heat superficial muscles and joints
- Speed resolution of hematomas
- Local hyperthermia in cancer patients

MICROWAVE CONTRAINDICATIONS

- General heat precautions
- Skeletal immaturity
- Because microwave diathermy selectively heats fluid-filled cavities, its use should be avoided in edematous tissue, moist skin, eyes, blisters, and fluid-filled cavities.
- Eye protection should be worn by the patient and the therapist due to risk of cataract formation.
- De Lateur et al. (1970) noted that the average temperature with microwaves at a depth of 1 to 3 cm is 105.8°F (41°C). At a frequency of 915 MHz, subcutaneous fat temperatures may increase by 10°C to 12°C. Muscles at a depth of 3 to 4 cm will be heated at only 3°C to 4°C.

Summary of Diathermy

- Diathermy produces deeper local temperature elevation within the tissues without overheating subcutaneous tissue or skin.
- All are forms of heating by conversion.
- It can be classified as:
  1. US
  2. SWD
  3. Microwave diathermy

ULTRASOUND	SHORTWAVE	MICROWAVE
• Sound waves • Frequency: 0.8–1.1 MHz • Heats at 8 cm depth (deepest penetration)	• Radio waves • Frequency: 27.12 MHz • Heats at 4–5 cm depth	• Microwaves • Frequency: 915–2,456 MHz • Superficial heat: 1–4 cm depth

INDICATIONS

ULTRASOUND	SHORTWAVE	MICROWAVE
• Chronic inflammation • Musculoskeletal pain • Contractures • Subacute trauma	• Chronic prostatitis • Refractory pelvic inflammatory disease • Myalgia • Back spasms	• Superficial heat for muscles and joints • Speeds the resolution of hematomas

CONTRAINDICATIONS

ULTRASOUND	SHORTWAVE	MICROWAVE
• General heat contraindications • Near heart and reproductive organs • Near tumors • Near brain, spinal cord, and laminectomy sites • Near pacemakers • Gravid or menstruating uterus • Infection sites • Skeletal immaturity • Total hip prostheses with methyl methacrylate	• General heat precautions • Metal • Contact lenses • Gravid or menstruating uterus • Skeletal immaturity	• General heat precautions • Skeletal immaturity • Avoid in fluid-filled cavities (eyes, blisters, moist skin, edematous tissue)

B. Cold

Therapeutic effects of cold are based on the following:

- Immediate local vasoconstriction
- Decrease in local metabolism
- Decrease in acute inflammatory response
- Slows nerve conduction velocity—decreased motor and sensory nerve conduction
- 🕮 Decreased spasticity due to:
  - Decreased muscle spindle activity—decreased firing rates of Ia and II afferent fibers
  - Decreased Golgi tendon organ activity—decreased firing rates of Ib afferent fibers
  - Research has evaluated the effect of using prolonged ice cooling (30 minutes) on the calf muscle and found a 3% to 10% decrease in elastic stiffness (Eldred et al., 1960; Lehmann & De Lateur 1990; Miglietta, 1973).
- Decreased pain/muscle spasm—increases nerve pain threshold
- Increased tissue viscosity with decreased tissue elasticity
- Transient increase in systolic and diastolic BP
- Release of vasoactive agents (histamine)

INDICATIONS FOR COLD THERAPY

- Generally used for acute processes:
  - Acute traumatic conditions—reduction of inflammation and edema in the 24- to 48-hour period
  - Immediate treatment of minor burns (via ice water immersion)
- Acute and chronic myofascial pain
- Musculoskeletal conditions and articular inflammatory states—arthritis, bursitis, and muscle strains/sprains
- Spasticity management

GENERAL PRECAUTIONS AND CONTRAINDICATIONS FOR COLD THERAPY

- Cold intolerance or hypersensitivity to cold (Raynaud disease/phenomenon)
- Arterial insufficiency—areas with circulatory compromise, such as ischemic areas in patients with peripheral arterial disease (PAD) affecting the arterial system
- Impaired sensation—insensate skin is at risk for burns
- Cognitive and communication deficits that preclude the patient from reporting pain
- Cardiac and respiratory involvement—if severe hypertension (HTN) is present, the patient's BP must be monitored closely.
- Cryotherapy-induced neuropraxia/axonotmesis, regenerating peripheral nerves
- Cryopathies: Cryoglobulinemia and paroxysmal cold hemoglobinuria
- Open wounds after 48 hours
- Note: Reflex vasodilation with hyperemia can occur after removal of ice.

MECHANISMS OF COLD TRANSFER

- Conduction: Cold packs and ice massage
- Convection: Cold baths (whirlpool)
- Evaporation: Vapocoolant spray
- The treatment modality depends on the size of the area to be treated and how accessible it is for cold application

CONDUCTION AGENTS

Cold packs:

- Include ice packs, wraps and sluices, endothermic chemical gel packs, and hydrocollator packs.
- The pack is wrapped in moist towels and treatment time is generally 15 to 20 minutes.
- Surface skin temperature can decrease by 15°C after 10 minutes; subcutaneous temperatures decrease by 3°C to 5°C.

- A study by Knutsson and Mattsson in 1969 showed muscle cooling by 5°C at a depth of 2 cm after 20-minute application of a hydrocollator pack.

Ice massage:
- For cooling of small areas (muscle belly, tendon, trigger point) before applying deep pressure massage.
- Combines the therapeutic effect of ice with the mechanical effects of massage.
- Direct application of ice to a painful area using gentle stroking motion.
- A study by Lowdon and Moore in 1975 showed a reduction of intramuscular temperature by 4.1°C at 2 cm depth in the posterior thigh region and up to 15.9°C reduction in biceps brachii after a 5-minute application time.
- Treatment of analgesia can be obtained in 7 to 10 minutes.

CONVECTION AGENTS

Cold baths:
- An example of hydrotherapy (i.e., whirlpool); uses water-filled containers for distal limb immersion
- Best suited for circumferential cooling of the limbs
- Water temperature: 4°C to 10°C
- Can be uncomfortable and poorly tolerated
- Effective for treatment of localized burns due to rapid skin temperature reduction

Evaporation:
- Vapocoolant sprays, such as Fluori-Methane spray, are less volatile and have widely replaced ethyl chloride sprays.
- 🕮 Used for spray-and-stretch techniques to treat myofascial and musculoskeletal pain (myofascial trigger point). It is also used for local anesthesia.
  - The technique consists of a series of unidirectional applications of vapocoolant sprayed parallel to the muscle fibers at a rate of 4 in./sec starting at the "trigger area" (area of deep myofascial hypersensitivity) and continuing to the "reference zone" (area of referred pain), while the muscle is passively stretched (Lavelle et al., 2007; Weber & Hoppe, 2011).
- Produces an abrupt temperature change over a small surface area.
- Precautions: Risk for skin site irritation and local cutaneous freezing.

Cold compression units:
- Combine the benefits of cold with the advantages of pneumatic compression
- Edematous extremities placed inside the sleeves with circulating cold water attached to an intermittent pump unit
- Used primarily to treat acute musculoskeletal injury with soft tissue swelling; also used after some surgical procedures
- Temperature: 45°F (7.2°C)
- Pressure: Up to 60 mmHg

LIGHT THERAPY

Ultraviolet Radiation

- Wavelength of 2,000 to 4,000 Å; bactericidal wavelength 2,537 Å
- Can be produced by a small, handheld mercury or "cold quartz" lamp
- Produces a nonthermal photochemical reaction with resultant alteration of DNA and cell proteins
- **Physiologic effects:**
  - Bactericidal on motile bacteria
  - 🕮 Increased vascularization of wound margins
  - Hyperplasia and exfoliation
  - Increased vitamin D production
  - Excitation of calcium metabolism
  - Tanning

- **Indications:**
  - For treatment of aseptic and septic wounds
  - 🕮 Psoriasis treatment, utilizing Goeckerman's technique, where a coal–tar ointment is applied to the skin prior to UV treatment
  - Acne treatment
  - Treatment of folliculitis
- **Precautions:**
  - Fair skin
  - 🕮 Scars and atrophic skin
  - Acute renal and hepatic failure
  - Severe diabetes
  - Hyperthyroidism
  - Generalized dermatitis
  - Advanced arteriosclerosis
  - Active, progressive pulmonary tuberculosis
  - Protect eyes from conjunctivitis, photokeratitis—shield from UV rays using goggles.
- **Contraindications:**
  - Pellagra
  - Porphyria
  - Sarcoidosis
  - Acute psoriasis
  - Lupus
  - Eczema
  - Herpes simplex
  - Xeroderma pigmentosum
- 🕮 Dosage is prescribed as the minimal exposure time required to cause erythema on the volar surface of the forearm; measured in minimal erythema dosage (MED), which subsides in 24 hours; usual initial prescription is in the dose of 1 to 2 MED and kept to <5
  - MED: Exposure produces a second-degree erythema in 4 to 6 hours with pain and subsides in 2 to 4 days, followed by desquamation.
  - 5 MED: Third-degree erythema in 2 to 4 hours with local edema and pain, followed by local desquamation.
  - 10 MED: Fourth-degree erythema with superficial blister.
- Treatment given two to three times a week

ELECTROTHERAPY

- Electrotherapy refers to the use of electricity to stimulate a peripheral nerve or muscle transcutaneously using electrodes.
- **Physiologic effects:**
  - Increases joint ROM
  - Muscle group contraction
  - Increases muscle strength
  - Increases circulation
  - Decreases muscle spasm
  - Slows muscle atrophy
  - Decreases spasticity
  - Promotes wound healing
  - Releases polypeptides and neurotransmitters (beta-endorphins, dopamine, enkephalins, vasoactive intestinal peptide, serotonin)
  - Induces osteogenesis—tissue regeneration and remodeling
  - Inhibits pain fibers—stimulates large myelinated Type A nerve fibers (Gate Control Theory; please refer to Transcutaneous Nerve Stimulation section)
  - Drives medicated ions across the skin

- **Indications:**
  - Pain management: Acute and chronic musculoskeletal pain, chronic neurogenic pain, and general systemic pain
  - Joint effusion and interstitial edema (acute and chronic)
  - Muscle disuse atrophy
  - Dermal ulcers and wounds
  - Circulatory disorders, including neurovascular disorders and venous insufficiency
  - Postherpetic neuralgia
  - Arthritis, including osteoarthritis and RA
  - ROM and stretching exercises
- **Contraindications:**
  - Circulatory impairment: Arterial or venous thrombosis and thrombophlebitis
  - Stimulation over the carotid sinus
  - Stimulation across the heart, especially if patient has pacemaker
  - Pregnancy
  - Seizure disorder
  - Fresh fracture
  - Active hemorrhage
  - Malignancy
  - Decreased sensation—direct current can cause electrical burns
  - Atrophic skin
  - Patient's inability to report stimulation-induced pain
  - Known allergies to gel or pads

TYPES OF ELECTROTHERAPY

Clinical uses of electrotherapy include pain management, muscle stimulation/reeducation, and medication delivery. The main types of peripheral electrotherapy that will be discussed are:

1. Transcutaneous nerve stimulation (TENS)
2. Neuromuscular electrical stimulation (NMES)
3. Iontophoresis

1. Transcutaneous Nerve Stimulation

A TENS unit uses a pocket-sized programmable device to apply a low-voltage electrical current through lead wires and electrodes attached to the patient's skin. It stimulates large-diameter sensory fibers to provide symptomatic relief of pain. An estimated 250,000 units are prescribed annually in the United States (Staats & Wallace, 2015).

- Electrode placement is subjective:
  - Typically placed over peripheral nerve distribution.
  - Locations can be distal or proximal to pain site.
- **Proposed mechanisms of pain control:**
  a. 🕮 Placebo effect is 30% to 35%.
  b. Believed to change an individual's perception of pain (Staats & Wallace, 2015).
  c. 🕮 The Gate Control Theory by Melzack and Wall (1965; **Figure 8–1**):
     - Attempts to account for mechanisms by which non-noxious stimuli can modulate pain sensation; that is, nonpainful stimuli can suppress pain.
     - TENS stimulates large IA (A-beta) myelinated afferent nerve fibers that in turn stimulate the substantia gelatinosa in the spinal cord, thus "closing the gate" on pain transmission to Lissauer's tract (posterior lateral tract of pain and temperature) and ultimately to the thalamus. As a result, pain signals can be blocked at the dorsal horn of the spinal cord before they are transmitted to the brain (Staats & Wallace, 2015).

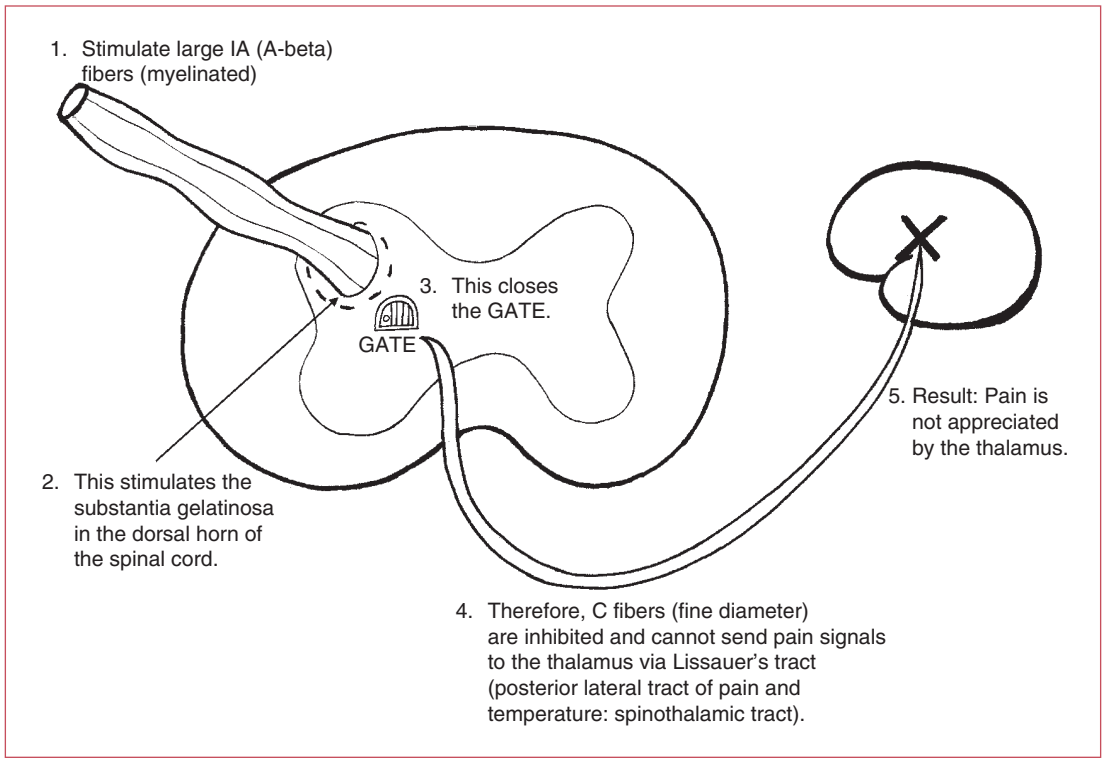

FIGURE 8–1 Simplistic visualization of the Gate Control Theory as described by Melzack and Wall (1965). Pain signals can be blocked at the level of the spinal cord before they are transmitted to the thalamus. (Follow steps 1–5 in the figure.)

d. Use of TENS releases endogenous opioids:
   - Cheng and Pomeranz (1979) demonstrated that pain relief produced at 4-Hz stimulation (low frequency) was blocked by naloxone; pain relief induced at 200 Hz was not blocked by naloxone. High-frequency stim versus low-frequency stim will activate delta versus mu, respectively. Opioid-tolerant individuals would benefit from high-frequency stimulation due to opioid analgesics' preference for mu-receptors (Léonard et al., 2011).

- Treatment time with TENS is normally 30 minutes to 1 hour per session, with a maximum of 2 hours per session, for a total of 8 hours per day. The treatments are continued for 3 weeks and gradually reduced over 8 to 12 weeks.
- 🕮 Studies on TENS revealed benefits in patients with:
  - Chronic low back pain—trend to greater pain reduction, better functional status, and better patient satisfaction versus placebo
  - Spinal cord injuries (Crane, 2014; Lin et al., 2010)
    - Effective in patients who experience mechanical/musculoskeletal or radicular pain at the level of injury
    - Less effective if applied below the site of injury
  - Osteoarthritic knee pain
  - Spastic paraspinal muscles—can be used in adjunct with myofascial trigger point injections
  - Labor pain
  - Primary dysmenorrhea, with high-frequency stimulation
  - Post amputation pain
  - Post mastectomy pain syndrome

- Overall, minimal side effects with long term-use. Patients may report discomfort or skin irritation if the intensity is too high. Skin irritation can be resolved if the electrode positions are shifted or if a different conducting gel is used. Electrode shifting can increase current intensities to uncomfortable levels.
- Tolerance may occur in the first few months with loss of efficacy.

TYPES OF STIMULATOR TECHNIQUES (TABLE 8–1)

CONVENTIONAL

- 🕮 High-frequency, low-intensity stimulation—most effective type of stimulation
- Thought to "close the gate" and inhibit nociceptive input at the interneuron level of the dorsal horn
- Duration of treatment: 30 minutes to hours
- Amplitude is adjusted to produce minimal sensory discomfort
- Pain relief begins in 10 to 15 minutes and stops shortly after removing stimulation
- Useful for neuropathic pain

TABLE 8–1 Types of Stimulator Techniques

TYPE	FREQUENCY	WIDTH (MSEC)	AMPLITUDE
Conventional	50–100	<200	Low
Acupuncture	1–10	200–300	High
Hyperstimulation	50–150	100–200	High
Burst	50–100[a] 1–10[b]	75–100	High
Modulated	Variable	<200	Variable

[a]Carrier frequency.
[b]Burst frequency.
Source: Adapted from Weber D, Brown AW. Physical agent modalities. In: Braddom RL, ed. *Physical Medicine and Rehabilitation*. W.B. Saunders; 1996, with permission.

ACUPUNCTURE

- Low-frequency, high-intensity stimulation
- Duration: 30 to 60 minutes
- Amplitude high enough to produce muscle contraction
- Onset of pain relief can be delayed several hours
- Pain relief persists hours after removing stimulation
- Useful for acute musculoskeletal conditions

HYPERSTIMULATION

- High-frequency, high-intensity stimulation
- Duration: Rarely tolerated more than 15 to 30 minutes
- Considered to stimulate the C-fibers and A-delta fibers and to cause counterirritation

PULSE (BURST) MODE

- High-frequency stimulation bursts at low-frequency intervals
- Duration: 30 to 60 minutes
- Delayed onset of pain relief

MODULATED

- Impulses vary in intensity and frequency
- Attempts to avoid neuro-habituation

2. Neuromuscular Electrical Stimulation

NMES refers to the process of applying electrical stimulation above the motor threshold to cause a muscle contraction. Successful use of NMES requires the alpha motor neuron to be intact. NMES systems utilize either external (most common) or internal electrodes to stimulate the muscle. NMES can be utilized as therapeutic muscle stimulation or for functional electrical stimulation (FES) as described here.

- System types:
  1. External (most common): Transcutaneous (surface) electrodes
     - Typical external systems use stimulation frequencies of 10 to 50 Hz.
  2. Internal: Implantable systems that use percutaneous, intramuscular, epineural, intraneural, and intraspinal electrodes

THERAPEUTIC VERSUS FUNCTIONAL ELECTRICAL STIMULATION

THERAPEUTIC NEUROMUSCULAR ELECTRICAL STIMULATION

- Repetitive stimulation is applied to paralyzed muscles to minimize atrophy and/or maintain ROM.

FUNCTIONAL ELECTRICAL STIMULATION

- Stimulation occurs in a coordinated sequence to assist the patient in performing a functional task, such as activities of daily living, transfers, or ambulation. FES systems employ either an open loop or a closed loop system.
- Open loop systems:
  - Feedback is provided manually.
  - Stimulation is activated by switches.
  - Intensity is adjusted based on response.
  - Example: Therapist triggers a heel switch to activate the anterior tibialis during gait cycle.
- Closed loop systems:
  - **Functional neuromuscular stimulation (FNS):** Term typically used to describe FES using these system types. FES and FNS are often used interchangeably (Chae et al., 2005).
  - More sophisticated system utilizing more complex automated technology.
  - Electrodes are activated by computer-generated patterns of stimulation to cause functional movement.
  - Feedback is provided automatically through movement sensors.
  - Electromyography (**EMG)–FES:** Surface EMG sensor triggers FES once threshold is reached; FES triggers desired movement.
  - Devices or systems that provide FNS are called neuroprostheses.
  - Examples:
    - Devices for foot drop: NESS L300, WalkAide
    - Devices used for upper limb function: NESS H200, Freehand System
    - Device for standing and gait: Parastep System

CLINICAL USES OF NEUROMUSCULAR ELECTRICAL STIMULATION

- 🕮 Strengthens muscles and maintains muscle mass after immobilization
- Provides feedback to enhance voluntary muscle control (neuromuscular reeducation)
- Provides cardiovascular conditioning (e.g., FES cycle ergometer in spinal cord injury [SCI])
- Prevents complications from immobility, such as deep vein thrombosis (DVT), disuse atrophy, and osteoporosis
- Shoulder subluxation in hemiplegic limb:
  - Baker and Parker (1986) published positive results of a protocol that provided external stimulation to the posterior deltoid and supraspinatus muscles. Similar protocols have since verified these findings (Chae et al., 2005).
- Spasticity management:
  - Stimulating spastic muscles to cause fatigue
  - Stimulation to antagonist muscle to produce reflex inhibition

  - Enhanced responses seen when used in conjunction with botulinum toxin injections and intrathecal baclofen (ITB) therapy
- Specific systems for phrenic nerve pacing and urinary incontinence

📖 *BENEFITS*

- Has been shown to potentially increase muscle mass, stroke volume (SV), and cardiac output (CO), as well as reduce venous pooling; also shown to potentially improve cardiorespiratory fitness (Bryce et al., 2011)

SIDE EFFECTS

- Allergic reaction of skin to electrode pads or gel
- Skin irritation and skin infection

PRECAUTIONS

- Avoid stimulation over the heart, neck, malignancies, pregnant uterus, or infected areas.
- May interfere with pacemakers.
- Caution with insensate skin (may cause burns).
- Caution with patients with seizure disorder.
- Avoid weakness from overworking muscle (use is controversial in neuromuscular diseases).
- Important to monitor BP when FES is used to exercise the lower extremities of SCI patients (especially patients over 45 years old or patients with a history of cardiovascular disease; Ragnarsson, 1988; Ragnarsson et al., 1988).

3. Iontophoresis

A transdermal delivery system in which a substance bearing a charge is propelled through the skin by a low electrical current. This method can be used to drive a drug across the skin barrier.

- **Uses:** Application of anti-inflammatory medication for conditions such as plantar fasciitis, tendinitis, and bursitis

THERAPEUTIC MASSAGE

Pressure and stretching are provided in a rhythmic fashion to the soft tissues. Treatments can be provided to the extremities for 5 to 15 minutes, or given to the trunk (neck, back, abdomen) for 15 to 30 minutes.

📖 Physiologic Effects

REFLEXIVE EFFECTS

- 📖 Reflex vasodilation with improvement in circulation
- Decrease in pain by means of the gate control or release of endogenous opiates or:
  - Neurotransmitters
  - General relaxation
  - Increased perspiration

MECHANICAL EFFECTS

- 📖 Assist in venous blood return from the periphery
- Increases lymphatic drainage
- Decreases muscle tightness
- Prevents or breaks adhesions in muscles, tendons, and ligaments
- Softens scars
- Loosens secretions—for example, chronic obstructive pulmonary disease (COPD)

Psychological Effects

- "Laying of hands" promotes a sense of general well-being.
- There is no effect on the metabolism. Massage will not affect muscle strength, mass, or rate of atrophy of denervated muscle.

Contraindications

- Do not perform over malignancies
- Avoid open wounds, infected tissues, or burns
- Nerve entrapments; severe pressure over trigger points has produced hematoma formation with subsequent nerve entrapment, in severe cases
- Acute inflammatory conditions: Gout, RA, cellulitis, and thrombophlebitis
- DVT
- Severe varicose veins
- Patients with severe clotting disorders or on anticoagulation

🕮 Common Techniques of Classic Therapeutic Massage

- **Effleurage**: Gliding, rhythmic strokes of the hand over the skin in a distal to proximal direction; performing gently and slowly results in muscle relaxation, whereas performing at a faster pace will increase stimulation. Deeper strokes result in more mechanical effects on the circulatory and deep myofascial system: relieving pain, increasing lymphatic drainage, and reducing vascular congestion associated with specific conditions (Brault et al., 2011).
- **Pétrissage**: "Kneading" technique in which the muscle tissue is "pinched" with the fingers and lifted from its underlying origin; it increases circulation and tissue pliability, and reduces edema and adhesions. Other variations include wringing, rolling, or shaking techniques (Brault et al., 2011).
- **Tapotement**: Percussion. Helps with desensitization, allows clearing of secretions, and improves circulation. Used for chest therapy in conjunction with postural drainage.
- **Friction massage**: Prevents adhesions in acute muscle injuries and breaks adhesions in subacute and chronic injuries. Also reduces local muscle spasm and decreases edema. Can be applied transverse or perpendicular to the muscle, tendon, or ligament fibers.
- **Soft tissue mobilization**: Forceful massage of the fascia muscle system. Massage is done with the fascia muscle in a stretched position rather than relaxed or shortened. Used for reduction of contractures.
- **Myofascial release**: Prolonged light pressure is applied in specific directions of the fascia system to stretch focal areas of muscle or fascial tightness.
- **Acupressure**: Finger pressure is applied over trigger points or acupuncture points to decrease pain.

🕮 MANUAL THERAPY

Manual manipulation involves treating motion restrictions based on the concept of somatic dysfunctions to improve a patient's overall function. Techniques can be characterized as either direct or indirect, and are classified as soft tissue, articular, or specific joint mobilization techniques.

Direct Techniques

- **Thrust**: Impulse, high-velocity, low-amplitude, and final activating force is from the operator.
- **Articulation**: Low velocity, high amplitude
- **Muscle energy (direct isometric type):** Introduced by Fred Mitchell, Sr., muscle energy technique involves an instructed movement of the patient against a defined resistance by the practitioner. There are three classifications based on whether the amount of force applied was equal (isometric), less (isolytic), or greater (isotonic). Patient contraction is the final activating force.
- **Direct myofascial release**: Tissue is held in a loaded, stretched position until muscle is released (Brault et al., 2011; Steindler, 1973; Weber & Hoppe, 2011).

Indirect Techniques (Brault et al., 2011; Steindler, 1973; Weber & Hoppe, 2011)

- **Strain–counterstrain**: Counterstrain tender points are identified and the area of the body is moved in a direction of freer motion, usually the same direction as the original injury, until tenderness is eliminated.
- **Indirect balancing**: The affected area of the body is moved away from the restrictive barrier until tension is equal on all sides of the dysfunction.
- **Functional (balanced ligamentous tension)**
- **Indirect myofascial release**
- **Craniosacral therapy**

TRACTION

Spinal traction provides a pulling force to the cervical or lumbar spine. Traction can be achieved using manual technique, a pulley system, or an electrical motorized device.

Physiologic Effects

- 🕮 Vertebral joint distraction—elongation of the cervical spine of 2 to 20 mm; can be achieved with 25 lb or more of tractive force
- Reduction of compression and nerve root and disc irritation
- Reduction of pain, muscle spasm, and inflammation
- Loosening of adhesions in the dural sleeves

Absolute Contraindications (Brault et al., 2011; Weber & Hoppe, 2011)

- Malignancy in the region of the spine
- Osteopenia and osteoporosis
- Infectious process (osteomyelitis, diskitis)
- Inflammatory arthritis
- Fracture
- Pregnancy
- Cord compression
- Congenital spinal deformity
- Uncontrolled HTN
- Cardiovascular disease (especially carotid or vertebral artery disease)
- Increased pain

🕮 Contraindications to Cervical Spine Traction

- Cervical ligamentous instability—RA, Down syndrome, Marfan syndrome, achondroplasia, Ehlers–Danlos syndrome
- Infectious process of the spine
- Cervical spinal stenosis with significant cord compromise
- Atlantoaxial subluxation with spinal cord compromise
- Vertebrobasilar insufficiency

Contraindications to Lumbar Spine Traction

- Pregnancy
- Cauda equina compression
- Aortic aneurysms
- Restrictive lung disease
- Active peptic ulcer disease
- Hiatal hernia

Prescription Parameters for Spinal Traction

POSITIONING

- For cervical traction: The neck is flexed with the patient in a sitting or supine position. To relieve symptoms of nerve root compression, 20 degrees to 30 degrees of flexion optimally opens the intervertebral foramina.
- For lumbar traction: Hip and knees are flexed to 90 degrees while the patient is supine (most common position used). This position reduces lumbar lordosis, and the spine is relatively flexed, opening the intervertebral foramina.

📖 *AMOUNT OF TRACTION*

- Cervical spine: Distraction requires >25 lb. Weights >50 lb do not provide an additional advantage. For cervical radiculopathy, traction may use 25 lb with neck flexion described earlier.
- Lumbar spine: Posterior vertebral distraction >50 lb is needed; for anterior separation, >100 lb is needed.
- The effect of friction between the treatment table and the body should be counterbalanced before true traction in the spine is accomplished.
- A study by Judovich reported that a pull equal to about one-half of the weight of the body part treated is needed to overcome friction. For the lower body, this is approximately 26% of the total body weight. Another option may be the use of a split table, which eliminates the lower body segment friction (Judovich, 1955).
- Regardless of the effect of friction, another 25% or more of body weight is needed to cause vertebral separation.

Intermittent Versus Continuous Traction

- Intermittent traction provides a greater pull. It is used for distraction when neural foramina opening or retraction of herniated disc material is desired.
- Continuous traction is used for prolonged muscle stretch, such as in muscle relaxation.
- Overall duration is typically 20 minutes.

THERAPEUTIC EXERCISE

Therapeutic exercises are prescribed to improve flexibility, endurance, aerobic capacity, and strength.

STRENGTHENING EXERCISES

- Strengthening exercises are designed to increase the maximal force that a muscle or muscle group can generate.
- Strength is affected by several factors:
  - Type of muscle contraction (eccentric vs. concentric)
  - Speed of contraction
  - Cross-sectional size of the muscle
  - Length–tension relationship of the muscle
  - Recruitment of motor units

📖 Muscle Physiology (Figure 8–2)

- Skeletal muscle fibers contain hundreds to thousands of myofibrils, each subdivided into functional units of contraction called sarcomeres.
- The sarcomere is composed of contractile proteins, actin and myosin, that lie parallel to the axis of the fiber. Muscle shortening is produced by coordinated movement of the thin (actin) and thick (myosin) filaments within the myofibrils.

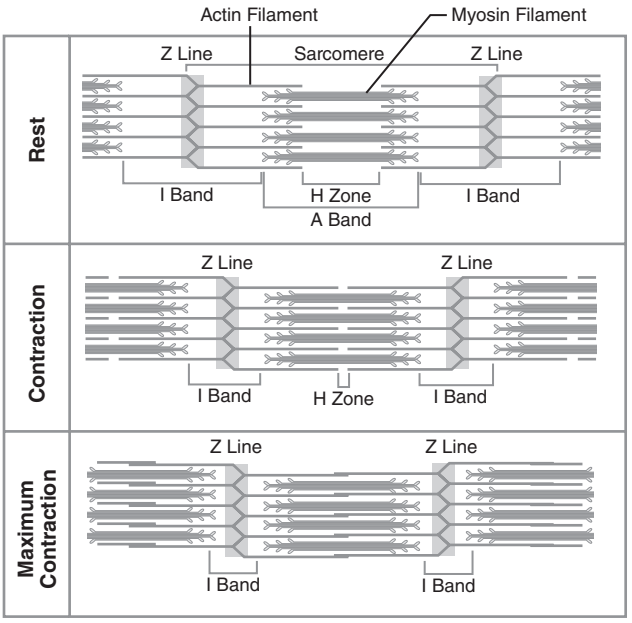

FIGURE 8–2 Sarcomere is represented schematically under different conditions: Rest, contraction, and maximum contraction. During contraction, the Z lines approach each other, and the H zone and I band shrink.

- The actin filaments attach to the outer margins of the sarcomere (Z line) and the myosin filaments are located centrally.
- The sarcomere is measured from Z line to Z line.
- During muscle rest, the filaments overlap:
  - The *A band* runs the length of the thick (myosin) filament. It has continuous overlap except at its center (*H zone*).
  - The *I band* is composed of thin (actin) filaments that remain bare through the outermost portion of the sarcomere.
- Thick and thin filaments are linked to one another via cross-bridges that arise from the myosin molecule. During muscle contraction, an increasing amount of myosin overlap is observed. Contraction results in the Z lines approaching each other, shrinking the H zone and the I band.
- Transient muscle fiber shortening takes place whenever an action potential is generated and travels through the sarcolemma (muscle fiber cell membrane).

SKELETAL MUSCLE FIBER TYPES (SEE ALSO CHAPTER 5, MUSCLE FIBER SECTION)

- Skeletal muscle fibers can be classified as Type I or Type II muscle fibers.

TYPE I MUSCLE FIBERS (SLOW-TWITCH, OXIDATIVE; RED)

- Used for low-intensity, long-duration activities
- Slower peak tension and relaxation
- Smaller muscle fiber diameter, innervated by slower conducting motor neurons
- High in oxidative enzymes, rich capillary supply

TYPE II MUSCLE FIBERS (WHITE)

- Used for high-intensity, short-duration activities
- Peak tension and relaxation achieved rapidly
- Large muscle fiber diameter, innervated by large, fast-conducting motor neurons
- Large number of muscle fibers (high innervation ratio)
- High activity of myofibrillar ATPase, which allows for high energy release for contraction
- High levels of glycogen and phosphorylase allow for high glycogen breakdown for anaerobic activities

- Divided into two subgroups:
  1. Type IIa: Fatigue-resistant (fast-twitch, oxidative/glycolytic); contains higher level of oxidative enzymes and better capillary supply than Type IIb
  2. Type IIb: Fast fatigable (fast-twitch, glycolytic)

Types of Strengthening Exercises

ISOTONIC	ISOMETRIC	ISOKINETIC
Muscle force generated with: • Visible joint movement • Variable speed • Constant external resistance (constant weight through ROM) Example: Weight lifting, DeLorme's PRE	Muscle force generated with: • No visible joint movement • Internal force does not overcome external force • Exertion against immovable objects or by holding joint in a static position Example: Isometric contractions done in bed	Muscle force generated with: • Visible joint movement • Constant speed • Variable external resistance Examples: Cybex, Nautilus

PRE, progressive resistive exercise; ROM, range of motion.

Open and Closed Kinetic Chain Exercises

- These are concepts in classifying strengthening exercises.
- **Closed kinetic chain (CKC) exercises** are activities in which the distal segment of a joint is fixed to an object, such as the ground (McGinty et al., 2000). An example of a CKC exercise is a squat or a push-up, where the foot/hand are "fixed" to the ground. CKC exercises tend to be more functional and more closely mimic the synergistic movements of everyday tasks by simultaneously activating both agonist and antagonist muscle groups (e.g., use of knee flexors and extensors during squats). They are initially better tolerated because they cause lesser shear forces and provide joint stabilization through muscle co-contractions (McGinty et al., 2000).
- **Open kinetic chain (OKC) exercises** are activities in which the distal segment of a joint is free to move and not stationary and fixed to an object. An example of an OKC exercise is a leg press (which moves a load away from the body using force) or bicep curl, where the foot/hand are not fixed to the ground. OKC exercises are optimal at strengthening specific muscle groups for a specific movement (e.g., isolated knee extension) and are therefore beneficial when correcting specific strength imbalances. Precaution must be taken since these exercises increase the shear forces in the soft tissue and at the intended joint, thereby potentially exacerbating certain conditions such as osteoarthritis or injuring the soft tissue (McGinty et al., 2000).

DELORME'S EXERCISES, ALSO KNOWN AS PROGRESSIVE RESISTIVE EXERCISES

- Dr. Delorme and Watkins (1948) developed a progressive resistive exercise (PRE) program to optimize a high-intensity resistance exercise program. It entails determining the greatest weight that can be lifted, pulled, or pushed 10 times through full ROM. The patient performs progressive sets of resistance exercises, with the patient lifting their 10 repetition maximum (RM). The patient initially lifts their 10 RM as follows: one set of repetitions at 50% of the 10 RM, a second set at 75% of the 10 RM, and a final set at 100% of the 10 RM. Each session consists of the three sets, with breaks between sets. The 10 RM is determined each week with progressive advancement as strength increases. PREs should be done at a moderate- to high-level intensity in order to be effective.
- A comprehensive exercise program prescription designed for strength training should include type of activity, frequency, intensity, duration, and precautions (Phillips & Kennedy 2012).

ECCENTRIC AND CONCENTRIC CONTRACTIONS

These are muscle contractions that may be isokinetic or isotonic.

ECCENTRIC CONTRACTIONS	CONCENTRIC CONTRACTIONS
• Lengthening contraction—resists a stretching force. • Fast eccentric contractions generate greatest amount of force. • Cause more tissue destruction • Increased muscle soreness up to 48 hours after initial muscle contraction. Can be minimized by beginning with low-intensity exercise, followed by regular exercise routine. • Decreased muscle soreness with muscle conditioning, which is best relieved by mild exercise of the affected muscle groups rather than rest • Low metabolic cost: Less VO_2 required (more energy-efficient)	• Muscle shortening against resistance; tension develops to overcome resistance • High metabolic cost • Generates little force

- Improvement of muscle performance, including strength, can be increased by:
  1. Increasing the amount of weight lifted
  2. Increasing the amount of repetitions or sets
  3. Increasing contraction velocity
- 🕮 **Figure 8–3** shows the relationship between force generation and velocity during eccentric, concentric, and isometric contractions.

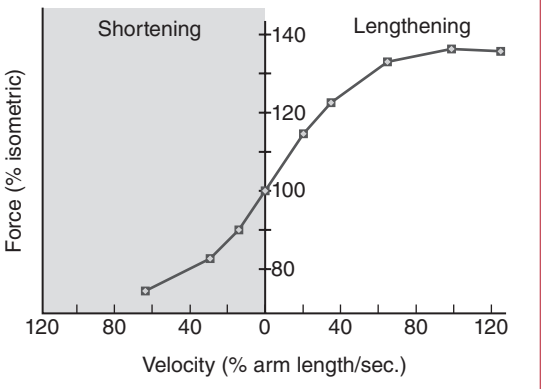

FIGURE 8–3 Greatest force is generated with fast eccentric contractions > slow eccentric contractions > isometric contractions > slow concentric contractions. Fast concentric contractions generate the least force.

🕮 Plyometric Exercises

- Training technique designed to increase muscular power and explosiveness (e.g., hopping, box jumps, forward hurdle hops).
- Should be used primarily as an advanced level of exercise in carefully selected patients, such as athletes who wish to return to high-demand functional activities. Exercises are intended to mimic motions used in sports.
- Uses short explosive movements that cause alternating eccentric and concentric muscle contractions. Increased elastic energy is stored in the eccentric phase, resulting in a more powerful concentric contraction.
- Due to high-intensity load on muscles, there is increased risk of injury.
- Appropriate only in the advanced phases of rehabilitation (Schache et al., 2009).

Anaerobic Exercises

- High-resistance, short-duration exercises at 80% to 100% of maximum exertion capacity
- Utilize the glycolytic system, which functions during the first 2 minutes of exercise and during high-intensity, short-duration exercising

Conditioning, Total Body Endurance Exercises, or Cardiopulmonary Endurance Exercises

AEROBIC EXERCISES (ENDURANCE EXERCISES)

These types of exercise use large muscle groups to perform continuous and rhythmic exercises that provide low intensity and high repetition. The goal with these aerobic exercises is to improve overall cardiopulmonary fitness. This is in contrast to anaerobic exercises, which utilize high-intensity and low-repetition routines.

- Combination of cardiopulmonary endurance exercise with strengthening
- Should consist of a warm-up period, a training period, and a cool-down period
  - Warm-up of 5 to 10 minutes
  - Training period of 20 to 30 minutes at 40% to 60% (low intensity), 60% to 70% (moderate intensity), or 70% to 85% (heavy intensity) of an individual's maximum oxygen consumption (VO_2 max)
  - Cool-down period of 5 to 10 minutes
- Guidelines for the quantity and quality of aerobic exercise programs for healthy adults per the American College of Sports Medicine and U.S. Department of Health and Human Services (Garber et al., 2011; Singh et al., 2020):
  - Method: Any exercise that uses large muscle groups, continuous and rhythmic in nature (e.g., running, swimming, walking, and stair climbing)
  - Frequency: 3 to 5 days a week
  - Moderate-intensity cardiorespiratory exercise training for ≥30 minutes, ≥5 days a week (total 150 minutes), or 75 minutes with vigorous-intensity activity with at least 2 days a week of muscle-strengthening activity
  - Goal of ≥500 to 1,000 metabolic equivalents (METs) per week
  - Flexibility exercises for each major muscle/tendon ≥2 days a week
  - Program tailored to an individual's physical ability and takes into consideration medical conditions and stated goals
  - Those who are unable or unwilling to meet the exercise targets, can still benefit from exercising less than the recommended amount

CARDIOVASCULAR EFFECTS OF CONDITIONING EXERCISES

- Decreased resting heart rate (HR) and submaximal effort
- Increased peak BP during maximal exercise; decreased BP at rest and submaximal effort
- Increased SV during maximal exercise
- Reduced myocardial oxygen consumption at rest and submaximal activities
- Comparison of cardiovascular effects on HR, VO_2 max, systolic and diastolic BP, and max VC, by exercise, can be found in **Table 8–2**.

Mobility Exercises

- Mobility exercises are designed to improve flexibility.
- Flexibility is defined as the ability to move body joints through their entire ROM.
- Each joint of the body has a specific ROM (**Table 8–3**).
- Flexibility exercises maintain mobility within the available ROM.
- Flexibility exercises should be done at least three times a week and should consist of three to five repetitions once or twice a day.

Exercise and the Effect on Cardiopulmonary Function (Table 8-2)

TABLE 8–2 Exercise and the Effect on Cardiopulmonary Function

EXERCISE	HR	VO_2 MAX	SBP	DBP	MAX VITAL CAPACITY
Aerobic	↑	↑	↑	↑	↑
Isometric	↑	No change	↑	↑	No change
Isotonic	↑	No change	↑	No change or ↓	No change
Effects after aerobic training	↓	↑	↓	No change or ↓	No change

DBP, diastolic blood pressure; HR, heart rate; SBP, systolic blood pressure; VO_2 max, maximum oxygen consumption.
Source: Savin WM, Alderman EL, Haskell WL, et al. Left ventricular response to isometric exercise in patients with denervated and innervated hearts. *Circulation*. 1980;61(5):897–901. doi:10.1161/01.CIR.61.5.897.

AVERAGE RANGE OF JOINT MOTION (IN DEGREES; TABLE 8–3)

TABLE 8–3 Average Range of Joint Motion (in Degrees)

SHOULDER	JOINT MOTION (DEGREES)	THUMB	JOINT MOTION (DEGREES)
Flexion	158°	Abduction	58°
Extension	53°	IP flexion	81°
Abduction	170°	MP flexion	53°
Adduction	50°	MC flexion	15°
Horizontal flexion	135°	IP extension	17°
Arm at side		MP extension	8°
Internal rotation	68°	MC extension	20°
External rotation	68°	**FINGERS**	**JOINT MOTION (DEGREES)**
Arm in 90-degree abduction		DIP flexion	80°
Internal rotation	70°	PIP flexion	100°
External rotation	90°	MCP flexion	90°
ELBOW	**JOINT MOTION (DEGREES)**	DIP extension	0°
Flexion	70°	PIP extension	0°
Hyperextension	90°	MCP extension	45°
FOREARM	**JOINT MOTION (DEGREES)**	**HINDFOOT**	**JOINT MOTION (DEGREES)**
Pronation	71°	Inversion	5°
Supination	84°	Eversion	5°
HIP	**JOINT MOTION (DEGREES)**	**FOREFOOT**	**JOINT MOTION (DEGREES)**
Flexion	113°	Inversion	33°
Extension	28°	Eversion	18°
Abduction	48°	**GREAT TOE**	**JOINT MOTION (DEGREES)**
Adduction	31°	IP flexion	60°
Horizontal flexion	60°	IP extension	0°
Hip in flexion		MTP flexion	37°
Internal rotation	45°	MTP extension	63°
External rotation	45°	**SECOND TO FIFTH TOES**	**JOINT MOTION (DEGREES)**
Hip in extension		DIP flexion	55°
Internal rotation	35°	PIP flexion	38°
External rotation	48°	MTP flexion	35°

(*continued*)

TABLE 8–3 Average Range of Joint Motion (in Degrees) (*continued*)

KNEE	JOINT MOTION (DEGREES)	Extension	40°
Flexion	134°	CERVICAL SPINE	JOINT MOTION (DEGREES)
Hyperextension	10°	Flexion	38°
ANKLE	JOINT MOTION (DEGREES)	Extension	38°
Plantar flexion	48°	Lateral bending	43°
Dorsiflexion	18°	Rotation	45°
WRIST	JOINT MOTION (DEGREES)	THORACIC AND LUMBAR SPINE	JOINT MOTION (DEGREES)
Extension	71°	Flexion	85°
Flexion	73°	Extension	30°
Ulnar deviation	33°	Lateral bending	28°
Radial deviation	19°	Rotation	38°

DIP, distal interphalangeal; IP, interphalangeal; MC, metacarpal; MCP, metacarpophalangeal; MP, metacarpophalangeal joint; MTP, metatarsophalangeal; PIP, proximal interphalangeal.
Source: From American Academy of Orthopaedic Surgeons. *Joint Motion: Method of Measuring and Recording*. AAOS; 1965, with permission.

Aquatic Exercises

- Pool-based therapy takes advantage of the buoyancy and viscosity of water.
- Due to buoyancy, the weight of a patient can be effectively reduced in proportion to water depth; a patient submerged in chest-deep water has a decreased weight-bearing load of 40% of total body weight.
- Viscosity of water allows for increased resistance to movement that is equal to the force exerted by the patient; resistance also decreases in proportion to the speed of movement.
- Studies have reported improvement in pain, fatigue, physical function, muscle relaxation, balance, coordination, and quality of life, with effects lasting up to 24 months after participation in pool-based therapy programs (Stanos et al., 2011).

Biofeedback

- Technique of using visual and audio equipment to reveal and translate normal and abnormal internal physiologic events in order to teach an individual to manipulate otherwise involuntary events. There are various types of biofeedback, each requiring its own set of equipment, setup, and testing. Biofeedback training requires that the patient have a potential for voluntary control, and the ability to follow commands and/or receptive aphasia.
- Clinical applications of biofeedback include stroke and spinal cord injury rehabilitation, traumatic head injuries, chronic pain management, urinary and fecal incontinence management, and sports-related muscle strengthening and physical training. Biofeedback has also been used in the management of specific conditions such as cerebral palsy, multiple sclerosis, dystonia, dyskinesias, peripheral nerve denervation, and Raynaud's disease. It has also been shown to have high efficacy when used to treat repetitive strain injuries.
- Types of biofeedback:
  - EMG biofeedback
  - Pressure or force biofeedback

  - Position biofeedback
  - Temperature and peripheral blood flow biofeedback
  - BP biofeedback
  - Respiratory biofeedback
  - Sphincter control training
- Although EMG biofeedback has been studied and is used extensively in stroke rehabilitation, it has less efficacy when there are major sensory deficits, especially severe deficits in proprioception. Additionally, proprioceptive deficits, marked spasticity, and receptive aphasia correlate with lower functional improvements when using biofeedback training. This was more evident in upper limb training than in lower limb training (Basmajian, 2005).

TECHNIQUES TO IMPROVE FLEXIBILITY

Can be done through anatomic planes of motion, combined planes of motion (similar to peripheral neurofacilitation patterns), or functional teaching activities.

🕮 Stretching Exercises

- Increase ROM by lengthening the tendon and muscle beyond the available range.
- Include static stretching, static stretching with contraction of the antagonist muscle (reciprocal inhibition), static stretching with contraction of the agonist muscle, and ballistic stretching
  - Static stretching: The joint is moved to the end of the ROM where the position is slowly held for 5 to 60 seconds. Can be done actively or passively, and is considered a safe technique.
  - Reciprocal inhibition: The joint is also moved to the end of ROM, and this is followed by a symmetric contraction of the antagonist muscle group for 5 to 30 seconds.
  - Static stretching with contraction of the agonist: The joint is moved to the end of ROM and followed by an isometric contraction of the agonist muscle for 5 to 30 seconds.
  - Ballistic stretching: Utilizes repetitive bouncing movements with a rapid stretch. More tension is developed and more energy is absorbed into the muscle and tendon, which can lead to bone avulsion or muscle–tendon tears. High risk of injury.

HAMSTRING INJURIES

- Hamstring injuries are commonly graded based on the degree of pain, weakness, and loss of ROM.
- The size and location of the hamstring injury are correlated with the amount of time required before returning to sports. The closer the injury is to the ischial tuberosity and if the hamstring tendon is involved, the longer the amount of time required prior to return to play.

HAMSTRING INJURY GRADING

- **Grade 0:** No damage
- **Grade I**: Tearing of a few muscle or tendon fibers
  - Minimal to no loss of strength.
  - MRI demonstrates minimal muscle/tendon injury (<5% of muscle length involved).
- **Grade II**: Partial tear of the musculotendinous unit but without complete disruption
  - Loss of hamstring strength.
  - MRI shows partial rupture (5%–50% of muscle length involved).
- **Grade III**: Complete hamstring muscle/tendon tear or avulsion fracture
  - Total loss of hamstring muscle function/strength, commonly associated with large hematoma.
  - Often requires surgery.

REHABILITATION PROTOCOL FOR HAMSTRING INJURIES (Clanton & Coupe, 1998)

- In the **acute phase** (up to 5 days), patients may start pain-free passive range of motion (PROM) after 24 hours from injury or active assistive range of motion (AAROM).
- In the **subacute phase** (day 3 to more than 3 weeks), patients may add on pain-free submaximal isometrics, stationary bike, and pain-free pool activities.

- In the **remodeling phase** (1–6 weeks), the patient may start prone eccentric exercises and apply moist heat or exercise prior to pelvic-tilt hamstring stretches.
- In the **functional phase** (2 weeks to 6 months) patient may start prone concentric exercises in addition to eccentric exercises and start pelvic-tilt hamstring stretching. Patient may start to jog, sprint, and perform sport-specific skills and drills.
- An athlete may return to full play when pain fully resolves, there is full ROM, and full strength returns.
  - Training programs should be progressive and include mainly core stabilizing exercises. Eccentric hamstring exercises, such as Nordic hamstring strengthening, have been shown to significantly reduce the number of hamstring injuries in both rugby and soccer players (Petersen et al., 2011).

EFFECTS OF EXTENDED BEDREST: IMMOBILIZATION AND INACTIVITY

MUSCLE

- 🕮 Immobilization decreases strength by 1.0% to 1.5% per day. Strength can decrease as much as 20% to 30% during only a week to 9 days of bedrest. Five weeks of total inactivity costs 50% of the previous muscle strength. A plateau is reached at 25% to 40% of original strength. One contraction a day at 50% of maximal strength is enough to prevent this decrease.
- 🕮 Percentage of muscle mass lost per week is estimated at 5% to 10%.
- Loss in muscle strength, especially in the quadriceps and extensors.
- A decrease in fiber diameter is found to affect type I fibers (slow-twitch) during early immobilization. A decrease in the percentage of Type I fibers is noted. A decrease in oxidative enzymes is also noted. All these changes also lead to a reduction in muscular endurance.
- Muscle torque can be affected: In a study by Gogia et al. (1988), the effect of bedrest on extremity muscle torque in healthy men was evaluated after they were strapped down for 5 weeks and allowed to sit only for bowel movements. A 26% decrease in gastrocnemius and 24% decrease in soleus muscle torque were found.
- Restricted activities affect muscle strength and recruitment patterns of muscles distant from specific sites of injury. In studies by Beckman and Buchanan (1995) and Nicholas et al. (1976), hip musculature strength and recruitment pattern were affected for several months after ankle sprain.

BONE AND JOINTS

- 🕮 Lack of stress and tension on the bone through weight-bearing and muscle pull on the bone causes bone loss and can lead to osteopenia. As a result of osteopenia, hypercalcemia develops. Calcium is excreted in the urine and feces starting at 2 to 3 days after immobilization and peaks at 3 to 7 weeks. After activity is resumed, calcium levels remain high for 3 weeks, reaching normal values at 5 to 6 weeks.
- Comparing calcium with nitrogen and protein changes in the urine, calcium improves last. Nitrogen loss is 2 g/day. Loss begins at 5 to 6 days after immobilization and peaks in the second week. After activity is resumed, loss continues for 1 week and plateaus during the second week. Below normal values are obtained at 4 weeks and return to normal values in 6 weeks.
- Calcium excretion, in addition to phosphorus loss, provides an increased likelihood for fractures.
- Joints show a decrease in periarticular connective tissue extensibility after inactivity. Articular cartilage begins to deteriorate due to lack of nutritional support. The hyaline cartilage in synovial joints is not supplied by vascular blood flow. It depends on nutrition from the synovial fluid through loading and unloading of pressure.

- Ligaments undergo biochemical changes noted as early as 2 weeks after immobilization. In the case of surgically repaired ligaments, improvement in strength is affected by immobilization. Immobilization causes ligament strength to decrease, compliance to increase, and collagen degradation to increase.

🕮 CARDIAC

- Reduction in blood and plasma volumes.
- Redistribution of body fluids leads to postural hypotension. Venous blood pooling occurs in the legs. In addition, beta-adrenergic sympathetic activity is increased.
- 🕮 Cardiovascular efficiency is decreased: Patients develop an increased resting HR and decreased SV.
  - HR increases approximately 0.5 beats/minute/day, leading to immobilization tachycardia and abnormal HR with minimal or submaximal workloads.
  - SV decrease may reach 15% with 2 weeks of bedrest as a result of blood volume changes and venous pooling in the lower extremities.
  - There is also a decrease in maximal oxygen extraction (VO_2 max) that can occur as early as 3 to 5 days.
- Increased risk of thromboembolism secondary to a decrease in blood volume and increased coagulability.

PULMONARY

- Diminished diaphragmatic movement and chest expansion due to loss of strength of diaphragm and intercostal muscles, leading to impaired secretion clearance and ventilation
- Reduced cough and bronchial ciliary activity. Patients can develop hypostatic pneumonia
- Reduction in pulmonary function with decreased tidal and minute volumes, and decreased VC
- A-V shunting and regional changes in ventilation–perfusion occur.

GASTROINTESTINAL

- Decreased gastrointestinal motility leads to constipation and loss of appetite

GENITOURINARY

- Urinary stasis, leading to an increased risk of urolithiasis and urinary tract infections

INTEGUMENTARY

- Skin atrophy and development of pressure ulcers

EVALUATION OF FUNCTIONAL INDEPENDENCE

🕮 It is important to understand the differences between **impairment, disability, and handicap** (Barbotte et al., 2001; Shepard, 1982):

1. **Impairment:** Physical or psychological abnormality of a body structure or function, usually the manifestation of a disease or injury; may be a temporary or a permanent loss
   - Example: Paraplegia due to SCI
2. **Disability:** A restriction or inability to perform an activity in the manner or within the range considered normal for a human being, mostly resulting from an impairment
   - Example: Inability to ambulate due to paraplegia
3. **Handicap:** Inability to perform or fulfill a usual role/life activity as a result of the impairment and disability; can be considered a social and cultural consequence of an impairment or disability

– Example: Inability to work upstairs in a building without an elevator due to an inability to ambulate from the paraplegia

TERM →	DEFINITION →	EXAMPLE
Disease	Sickness	Radial nerve palsy
Impairment	Physical, anatomic, or psychological abnormality	Wrist drop
Disability	Loss of function	Inability to write
Handicap	Loss of role in society	Loss of job as an artist

- The evaluation of functional independence is important in the process of assessing a safe return home after a patient has been hospitalized.
- 🕮 The **Functional Independence Measure (FIM) score** is one of the common measurement scales used for this type of assessment **(Figure 8–4).**
  - It documents the severity of disability, measures activity limitations, and documents outcomes of rehabilitation treatment as part of a uniform data system.
  - Consists of 18 items organized under six categories:
    - Self-care (eating, grooming, bathing, upper body dressing, lower body dressing, and toileting)
    - Sphincter control (bowel and bladder function)
    - Transfers (bed, chair, wheelchair, tub or shower, and toilet transfers)
    - Locomotion (ambulation, wheelchair mobility, and stair management)
    - Communication (comprehension and expression)
    - Social cognition (interaction, problem-solving, and memory)
  - Patients are graded in each category on a scale of 1 (total assistance required) to 7 (complete independence).
- 🕮 FIM is an ordinal scale (see Chapter 12, Associated Topic).

PHYSIOLOGIC EFFECTS OF AGING

🕮 CARDIAC

- Progressive decline in maximal HR, possibly related to decreased chronotropic response to adrenergic stimuli. Note: There is no change to resting HR with aging.
  - Clinical formula for maximal HR:
    - Traditional calculation: Maximum HR = 220 – age
    - Note: While this calculation is used traditionally, there is literature that states this calculation is more applicable to the male population. Adjustments likely need to be made when calculating for females. This calculation appears to underestimate peak HR in women 50 to 90 years of age, and overestimate peak HR in women 40 to 50 years of age. A recent study suggests a more applicable calculation for females would be 210 – (0.79 × age). Since there is not yet a consensus on updated calculations, experts suggest using other markers to guide duration of exercise prescriptions (Sydó et al., 2014).
- Increased left ventricle end-systolic volume and decreased ejection fraction with exercise. When this is added to a decreased HR response, CO during exercise is more dependent on an increased SV by using the Frank–Starling mechanism (higher left ventricular end-diastolic volumes).
- CO decreases with aging, leading to an increased risk of congestive heart failure.

FIM® Instrument

LEVELS		
	7 Complete Independence (timely, safely) 6 Modified Independence (device)	NO HELPER
	Modified Dependence 5 Supervision (subject = 100%) 4 Minimal Assistance (subject = 75%+) 3 Moderate Assistance (subject = 50%+) **Complete Dependence** 2 Maximal Assistance (subject = 25%+) 1 Total Assistance (subject = less than 25%)	HELPER

	ADMISSION	DISCHARGE	FOLLOW-UP
Self-Care			
A. Eating			
B. Grooming			
C. Bathing			
D. Dressing: Upper Body			
E. Dressing: Lower Body			
F. Toileting			
Sphincter Control			
G. Bladder Management			
H. Bowel Management			
Transfers			
I. Bed, Chair, Wheelchair			
J. Toilet			
K. Tub, Shower			
Locomotion	W Walk C Wheelchair B Both	W Walk C Wheelchair B Both	W Walk C Wheelchair B Both
L. Walk, Wheelchair			
M. Stairs			
Motor Subtotal Rating			
Communication	A Auditory V Visual B Both V Vocal N Nonvocal B Both	A Auditory V Visual B Both V Vocal N Nonvocal B Both	A Auditory V Visual B Both V Vocal N Nonvocal B Both
N. Comprehension			
O. Expression			
Social Cognition			
P. Social Interaction			
Q. Problem Solving			
R. Memory			
Cognitive Subtotal Rating			
TOTAL FIM® RATING			

NOTE: Leave no blanks. Enter 1 if patient is not testable due to risk.

FIGURE 8–4 FIM score documents the severity of disability as well as the outcomes of rehabilitation treatment. *Source:* Copyright® 1997 Netsmart Technologies, Inc. Reprinted with permission of Netsmart, 11100 Nall Avenue, Overland Park, KS 66211.

- Rate of diastolic early filling is decreased. It is more dependent on late filling through atrial contraction.
- Patients are more susceptible to atrial fibrillation or atrial tachycardia, as well as congestive heart failure.

- Maximal oxygen consumption (VO_2 max) decreases regardless of the level of activity, but more physically active patients have smaller decreases compared with sedentary patients.
- Progressive, gradual increases in systolic and diastolic BP are mostly due to decreased arterial elasticity than to circulating catecholamines.
- Decreased baroreceptor sensitivity is associated with increased incidence of symptomatic orthostatic hypotension: Diminished reflex tachycardia with position change, in association with blunted plasma renin activity and reduced vasopressin and angiotensin II levels.
- Decreased baroreceptor sensitivity is also associated with cough and micturition syncope syndromes.
- The patient's medications should be monitored during evaluation of the causes of orthostatic hypotension: Antihypertensives, levodopa, phenothiazines, and tricyclic antidepressants (TCA). Also evaluate for decrease in fluid intake, fluid loss, and aldosterone/cortisol level dysfunction.

PULMONARY

- Progressive decline in pulmonary function occurs with aging, primarily due to loss of reserve capacity
- Decreased VC
- Decreased PO_2: Linear decline associated with a mild degree of impaired gas exchange
- No changes in PCO_2 or pH
- Oxygen saturation remains normal or slightly reduced
- Decreased forced expiratory volume in 1 second (FEV_1). This decreases consistently at 30 mL per year
- Decreased maximum minute ventilation. These declines reflect changes in related organ systems, which are stressed by voluntary ventilation required during these tests
- Examples: Stiffening of the rib cage, weakening of the intercostal and abdominal muscles, and small airway narrowing due to decreased elastic recoil (i.e., decreased compliance)
- Increase in residual volume and functional residual capacity related to loss of elastic recoil of the lung tissue
- No change in total lung capacity
- Progressive ventilation–perfusion imbalance due to collapse of small peripheral airways with decreased ventilation of alveoli; results in a linear decline in pO_2 with aging: $pO_2 = 110 - (0.4 \times \text{age})$
- pO_2 lower in supine position than with sitting or standing
- No changes in pCO_2 or pH
- O_2 saturation typically normal or only slightly reduced
- More vulnerable to significant hypoxia from minor insult due to position on oxygen hemoglobin dissociation curve
- Increase in sleep-related breathing disorders with aging
- High incidence of pneumonia: Immunologic decline, impaired mucociliary action, decreased chest wall compliance with decreased ability to clear secretions, reduced level of consciousness, dysphagia, and esophageal disorders

INTEGUMENTARY

- Decreased elasticity of connective tissue leads to increased risk of pressure ulcers and skin breakdown
- Decreased moisture
- Decreased skin sensation

MUSCULOSKELETAL

- Progressive loss of muscle strength with aging from the fourth decade due to overall decrease in muscle cross-sectional area and mass with age
- Sarcopenia refers to the age-related loss of muscle mass, muscle strength, and physical performance:
  - Overall muscle mass decreases, including the number of myofibrils and concentration of mitochondrial enzymes
  - Decreased number of motor units
  - Combined with obesity can lead to further reduction in function
- With endurance training, muscle endurance increases or remains stable due to muscle fiber type regrouping
- There is an increase in Type IIa fibers with a similar decrease in Type IIb fibers; can occur due to the fact that Type II fibers become more aerobic with endurance training
- With endurance training, the percentage of Type I muscle fibers does not change in either older or younger adults (Kirkendall & Garrett, 1998)
- Increased fat: 15% increase at age 30; 30% increase at age 80; patients retain more fat-soluble medications and have increased side effects
- High prevalence of osteoporosis and degenerative joint disease—decreased water content of cartilage and ratio of chondroitin-4-sulfate to chondroitin-6-sulfate; chemical alteration of cartilage with a decreased ability to bear weight without causing ulceration of cartilage and eventual exposure of bone
  - Wear and tear hypothesis of osteoarthritis—result of cumulative stresses of a lifetime of joint use
  - "Primary" osteoarthritis results from stress of repetitive weight loading or strain
  - "Secondary" osteoarthritis may be related to occupational or congenital factors with unusual patterns of stress
- Decreased peak bone mineral density starting in mid-30s, leading to concerns of osteoporosis
- Base of stance is increased
- Treatment includes adequate nutrition and targeted exercise

THERMOREGULATORY

- Impaired thermoregulation with decreased autonomic vasomotor control and impaired sensitivity to changes in temperature.
- Patients are vulnerable to hypothermia and hyperthermia. Hypothermia is affected by impaired thermogenesis and aggravated by medical conditions such as malnutrition, hypoglycemia, and hypothyroidism; or medications such as narcotics, ethanol, and benzodiazepines. Hyperthermia is affected by decreased sweat production, a major contributing factor in heat exhaustion and heat stroke in hot conditions. Hypohidrosis is aggravated by anticholinergics, phenothiazines, and antidepressants (Cameron et al., 2020).

NEUROLOGIC

- Older adults are capable of learning but at a slower rate:
  - Decreased short-term memory and incidental learning (Bolla et al., 1991).
  - Slowing in the rate of central information processing tasks requiring new information processing tends to decline progressively after 20 years of age.
- Increase in choice reaction time. The more complex the task, the greater the age effect (Bolla et al., 1990).
- Decrease in proprioception and gait, leading to problems with coordination and balance. There is an associated decrease in nigrostriatal neurons with age.
- Loss of speed of motor activities and decreased balance correction.

- In otherwise healthy individuals, there is a greater decline in the ability to perform mathematics as compared with language comprehension (Bleecker et al., 1988a). Results have shown that memory disturbances in older adults are related to depression rather than poor performance. Related studies show that there is little or no aging effect on verbal ability. This effect was found to be greater in men. Men had a lower overall verbal score than women in the 66- to 89-year-old age group.
- Dementia is the loss of cognitive functioning to an extent that it interferes with a person's daily life and activities.
  - Primary risk factor for development of dementia is age.
  - Dementia associated with aging includes Alzheimer disease (most common), frontotemporal disorders (e.g., behavioral variant frontotemporal dementia, primary progressive aphasia), Lewy body dementia, and vascular contributions.
  - 🕮 Reversible dementia-like conditions include normal pressure hydrocephalus, nutritional deficiencies (vitamin B_1 or B_{12}), adverse effects of medications, vasculitis, subdural hematomas, nonmalignant brain tumors, and chronic infections (Cameron et al., 2020).

GENITOURINARY

- Reduction in bladder capacity
- Decreased urethral and bladder compliance
- Reduced urinary flow rate
- Decreased ability to postpone voiding
- Incontinence does not result from normal aging. It is the result of underlying disease
  - Approximately 70% of older adult patients with urinary incontinence have detrusor instability.
  - Confusional states, urinary tract infections, atrophic urethritis, drugs (sedatives, anticholinergics, calcium channel blockers), limited mobility, and constipation can contribute.
- Sexuality: Decreased sexual function with aging
  - Older men have decreased ability to have psychogenic erections and require more intense stimulation. Erections may be partial; the force of ejaculation is decreased, with a less intense sensation of orgasm. Many medications can contribute to impotence.
  - Women undergo postmenopausal changes, such as increased vaginal wall fragility and decreased vaginal lubrication. Other reasons for decreased sexual function may include partner's impotence, decreased libido, and decreased opportunities for sexual encounters.
- Benign prostatic hyperplasia (BPH), which develops under hormonal influence and is almost universal in men over 40 years of age

RENAL

- Age-related decrease in renal function of about 1% per year from approximately 40 years of age.
  - Decrease in creatinine clearance of 7.5 to 10 mL per decade
  - No change in serum creatinine and blood urea nitrogen (BUN) levels
- Impaired ability to concentrate and dilute urine, impaired sodium conservation, reduction of urine acidification, and decreased ability to excrete an acid load.
  - More susceptible to electrolyte and fluid imbalances
  - Major concern for hyponatremia due to water intoxication, especially when complicated with an acute illness due to nonspecific signs including depression, confusion, lethargy, anorexia, and weakness (Cameron et al., 2020)
- Atrophy of the glomeruli and decrease in renal tubular cell mass, leading to decreased glomerular filtration rate and decreased tubular function.
- These factors result in an increase in the half-life of renally excreted drugs, such as penicillin, aminoglycosides, digoxin, cimetidine, lithium, procainamide, and chlorpropamide.
- 🕮 Digitalis toxicity in older adults is commonly secondary to impaired renal function. Toxicity manifests with cardiac dysrhythmias, anorexia, nausea, vomiting, abdominal pain, fatigue, depression, drowsiness, lethargy, headache, confusion, and ocular disturbances.

- 🕮 Nonsteroidal anti-inflammatory drugs (NSAIDs) can also cause injury to the kidneys, with the most common finding being prerenal azotemia. In settings where the renal blood flow is dependent on the activity of vasodilating prostaglandins, NSAIDs can precipitate acute renal failure. In states of prostaglandin inhibition, patients develop hyporeninemic hypoaldosteronism.

GASTROINTESTINAL

- 🕮 **Dysphagia** is a health concern in older adults. Age-related changes in swallowing physiology as well as age-related diseases are predisposing factors for dysphagia in older adults (Sura et al., 2012).
- Impaired esophageal function: Decreased amplitude in peristaltic contractions.
- Delayed esophageal emptying and incomplete sphincter relaxation are associated with disease.
- Colon: Decreased force of muscle contraction with impaired rectal perception of feces. It is important to explore the contributing factors to the development of constipation, including decreased dietary fiber and fluid intake, diseases associated with decreased bowel function (Parkinson, cerebral vascular accidents [CVA]), and medications (calcium, iron, antacids, NSAIDs, opiates, antihypertensives [calcium channel blockers], anticholinergics [TCAs], sympathomimetics [isoproterenol, terbutaline, pseudoephedrine]).
- Fecal incontinence is secondary to fecal impaction in most cases. Other causes, such as decreased cognitive function, diarrhea, and decreased sphincter tone, need to be evaluated. Diarrhea is commonly seen in association with fecal impaction, infection, and drugs such as laxatives, antibiotics, and digoxin toxicity.

HEARING

- 🕮 Presbycusis is the loss of ability to perceive or discriminate sounds as one ages. The age of onset and pattern may vary (Stedman, 2005).
- Most commonly manifested as a conductive hearing loss, possibly due to increased stiffness of the basilar membrane, increased threshold sensitivity, narrower range of audibility, abnormal loudness, and difficulty discriminating complex sounds (Cameron et al., 2020).

EFFECTS OF ACUTE HOSPITALIZATION AND DECONDITIONING IN OLDER ADULTS

- Multiple chronic illnesses have cumulative effects on the organ reserves, to which the older adult patient may try to adapt in order to be functional under the circumstances.
- The patient's reserve capacity may be already limited, and minor acute complications or disease processes may result in functional decompensation.
- Adverse effects of hospitalization in older adults include the following:
  - Disorientation
  - Insomnia—usually treated with hypnotics, which may cause side effects or may adversely affect the patient's health. Sleep-related disorders in older adults are frequently related to depression.
  - Increased incidence of iatrogenic complications—adverse drug interactions are usually a result of polypharmacy.
  - Emotional sequelae—anxiety and confusion are common in relation to illness, prognosis, and hospitalization. Patients tend to develop depression. Many times, patients are dependent for functional activities.
  - Social support system and discharge disposition—patient's functional abilities may be impaired. This can be added to a decrease in patient's motivation, which can lead to a more difficult return to a prior living situation.
- Deconditioning effects, as previously discussed, tend to appear earlier, are more severe, and take longer to reverse in older adults. This is due to age to a great degree and a diminished reserve. Deconditioning effects include:

  - Decreased VO_2 max
  - Shortened time to fatigue during submaximal work
  - Decreased muscle strength
  - Decreased reaction time/balance/flexibility
- Multiple factors associated with falls are secondary to deconditioning. Falls in the community are associated with decreased static and dynamic balance, leg strength, hip/ankle flexibility, vision deficits, and neuropathies.

SUMMARY OF ADAPTATIONS TO EXERCISE IN OLDER ADULTS

Aerobic Conditioning

- Decreased minute ventilation during submaximal exercise—decrease of 9% to 15%.
- HR decreases with submaximal exercise, with a decrease of 9 to 20 beats per minute, and SV increases 8%.
- Minute ventilation increases with maximal exercise—decrease 20% to 30%.
- With maximal exercise, SV and CO increase—SV 6% to 8%; CO up to 34%.
- Total hemoglobin (Hb) and blood volume increase—Hb 7%; blood volume 8%.
- Systemic vascular resistance during submaximal exercise—decreases 5% to 18%.
- Muscle enzymes increase oxidative enzymes—0% to 45%; glycogen stores 10% to 28%.

Strength Training

- Increase in muscle cross-sectional area, Type I and II fibers, capillary density per fiber, and oxidative enzymes
- Age-related decrease in sarcoplasmic reticulum Ca^{2+}—ATPase is prevented
- Strength and torque velocity increase.

MANAGEMENT OF COMPLICATIONS IN OLDER ADULTS

Depression

- Can present as sleep disturbance, loss of appetite, constipation, impaired concentration, poor memory, and psychomotor retardation. Rates of major depression vary from 16% to 30% in geriatric populations. Prevalence rates in community-dwelling older adults range from 2% to 5%. This risk is increased threefold if the patient has a disability.
- For prolonged depression, trazodone and selective serotonin reuptake inhibitors (low anticholinergic effect) are good. TCAs with the lowest anticholinergic effects can also be effective.
  - 🕮 Of the common TCAs, nortriptyline (Pamelor) is preferred over others such as amitriptyline. Nortriptyline has fewer anticholinergic effects, has a decreased sedation effect, and causes less orthostatic hypotension (which is a result of alpha-1-blockade).

🕮 Agitation

- If medications are required, amitriptyline (Elavil) is recommended over other medications such as benzodiazepines, including Valium. Due to the fact that fat-soluble drugs tend to accumulate more in older adults as a result of changes in metabolism and body composition, benzodiazepines accumulate, with adverse side effects including drowsiness.

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PULMONARY, CARDIAC, AND CANCER REHABILITATION

9

PULMONARY REHABILITATION—Priscila Gonzalez, MD • Nicholas G. Melillo, MD • Daphne Karen MacBruce, MD • Sara J. Cuccurullo, MD • Steven Markos, MD • Lisa Casale, MD • Phillip Gordon, MD

CARDIAC REHABILITATION—Iqbal Jafri, MD • Sara J. Cuccurullo, MD • Talya Fleming, MD • Steven Markos, MD • Stacey Jou, MD

CANCER REHABILITATION—Priscila Gonzalez, MD • Leslie Bagay, MD • Ofure Luke, MD • Anna Maria Dunn, MD • Lisa Luciano, DO • Richard M. Schuman, MD • Eric Heckelmann, MD

PULMONARY REHABILITATION

GOALS OF PULMONARY REHABILITATION

- Improvement in cardiopulmonary function
- Prevention and treatment of complications
- Increased understanding of the disease
- Increased patient responsibility for self-care, disease-specific self-management, and compliance with medical treatment
- Improvement in quality of life, capacity for activities of daily living (ADLs), and return to the community at a functional level as close to the prior level as possible

🕮 BENEFITS OF PULMONARY REHABILITATION

- Improvement in exercise tolerance, symptom-limited oxygen consumption, work output, mechanical efficiency
- 🕮 Exercise increases arterial venous oxygen (AVO_2) difference by increasing oxygen extraction from arterial circulation.
- Reduction in dyspnea and respiratory rate (RR) at rest and at various levels of activity
- Improvement in general quality of life, decreased anxiety and depression, as well as improvement in the capacity to perform ADLs
- Improvement in ambulation capacity and endurance
- Decreased hospitalization rates/reduced healthcare resource utilization
- Focus on conditioning peripheral musculature in order to improve efficiency and reduce stress on the heart and lungs (Alba, 1996)

CANDIDATES FOR PULMONARY REHABILITATION

- 🕮 Patients who benefit the most from a pulmonary rehabilitation program have at least one of the following:
  - Respiratory limitation of exercise at 75% of predicted maximum O_2 consumption
  - Obstructive airway disease with a forced expiratory volume in 1 second (FEV_1) <2,000 mL or an FEV_1/FVC (forced vital capacity) ratio <60% (see Lung Volume Definitions section and **Figure 9–1** for further details)
  - Restrictive lung disease or pulmonary vascular disease with carbon monoxide diffusion capacity <80% of predicted value (Bach, 1993)
- Motivated nonsmokers or patients who have quit smoking and whose activities are limited because of dyspnea are good candidates for a pulmonary rehabilitation program. Cigarette smokers can benefit from rehabilitation if the role of smoking cessation is emphasized and integrated into the rehabilitation program (Celli & ZuWallack, 2005).
  - Functional evaluation to assess pulmonary disability is recommended prior to starting the program and includes pulmonary function testing (PFT) and assessment of exercise capacity with a 6-minute walk test, shuttle test, or cardiopulmonary stress test.
- The 2024 Global Initiative for Chronic Obstructive Lung Disease (GOLD) Guidelines encourages patients with a high symptom burden and increased risk of exacerbation (GOLD Groups B and E) to participate in a formal pulmonary rehab program (**Figure 9–1**).

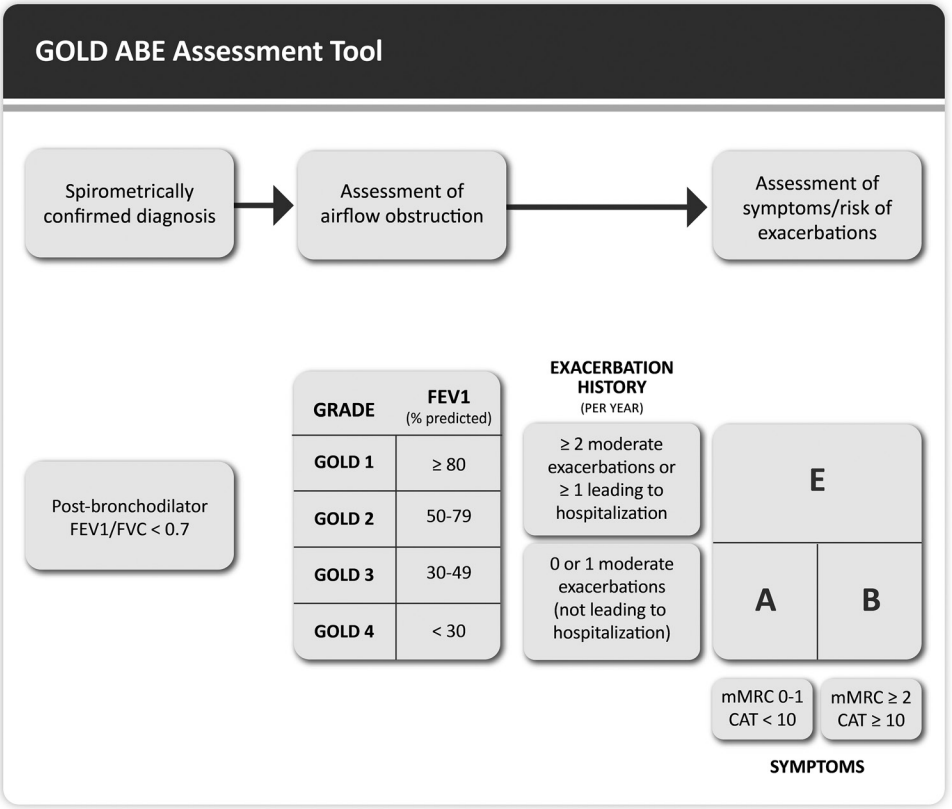


FIGURE 9–1 GOLD ABE Assessment Tool proposed by GOLD in 2023.
Note: A,B,E are patient groups (see GOLD Guidelines for further description). The 2024 GOLD Guidelines encourages patients with a high symptom burden and increased risk of exacerbation (Gold Groups B and E) to participate in a formal pulmonary rehab program.
CAT, COPD Assessment Test; FEV1, forced expiratory volume in 1 second; FVC, forced vital capacity; mMRC, modified Medical Research Council dyspnea scale (dyspnea questionnaire).
Source: From Global Initiative for Chronic Obstructive Lung Disease. Global strategy for the diagnosis, management, and prevention of chronic obstructive pulmonary disease: 2024 report. https://goldcopd.org/2024-gold-report.

- Contraindications to participation in pulmonary rehabilitation include angina pectoris, recent myocardial infarction (MI), severe pulmonary hypertension (pHTN), decompensated congestive health failure, acute cor pulmonale, severe exercise-induced hypoxemia not correctable with supplemental oxygen, inability to perform exercises due to severe orthopedic impairments, and severe cognitive deficits (Sharma & Singh, 2011).

🕮 CLASSIFICATION OF FUNCTIONAL PULMONARY DISABILITY: MOSER CLASSIFICATION

(Corsello, 1991)

1. Normal at rest—dyspnea on strenuous exertion
2. Normal ADL performance—dyspnea on stairs/inclines
3. Dyspnea with certain ADLs; able to walk one block at slow pace
4. Dependent with some ADLs; dyspnea with minimal exertion
   Note: 1 to 4 have no dyspnea at rest.
5. Housebound—🕮 dyspnea at rest, in need of assistance with most ADLs
   - Though patients with the criteria listed earlier may benefit from rehabilitation, there are no minimum requirements of lung function to include patients in pulmonary rehabilitation (Celli & ZuWallack, 2005).
   - The GOLD Guidelines is used to classify patients based on exacerbations and symptom score questionnaires (GOLD 2024 Report).

REVIEW OF PULMONARY PHYSIOLOGY

Control of Respiratory Function

- Voluntary control of respiration originates in the medulla oblongata and descends through the spinal cord to the respiratory muscles.
- The medullary respiratory center serves to integrate different chemoreceptors (Axen, 1991).
  - Central chemoreceptors are stimulated by hypercarbia in the cerebrospinal fluid.
  - Peripheral chemoreceptors in the carotid and aortic bodies are stimulated by alterations in the carbon dioxide, oxygen, and pH levels in the blood.

Muscles of Respiration

ACTIVE MUSCLES DURING INSPIRATION

- 🕮 The diaphragm, innervated by the phrenic nerve, is the primary muscle of respiration during inspiration. Diaphragmatic contraction increases the volume and decreases the intrathoracic pressure in the thoracic cavity. This resulting decrease in intrathoracic pressure relative to atmospheric pressure results in inhalation.
- Accessory muscles of respiration, primarily of use during exertion or respiratory distress, include the sternocleidomastoid, trapezius, pectoralis major, external intercostals, and scalene muscles.

ACTIVE MUSCLES DURING ACTIVE EXPIRATION

- Normally expiration is a passive process but becomes more active during exercise and in certain disease states, such as emphysema.
- Abdominal muscles—primary expiratory muscles
- Accessory muscles: Internal intercostals

ACTIVE MUSCLES OF THE UPPER AIRWAY

- Facilitate airway patency
- Include muscles of the mouth, tongue, uvula, palate, and larynx
- Significant relaxation of these muscles can lead to airway obstruction during sleep, as seen in obstructive sleep apnea (OSA).

Causes of Acute Respiratory Failure

- Severe respiratory infections
- Pulmonary edema
- Exacerbations of chronic lung disease such as asthma, chronic obstructive pulmonary disease (COPD), or interstitial lung disease
- Diffuse parenchymal injury or lung disease
- Acute respiratory distress syndrome (ARDS)
- Acute pulmonary circulatory failure (e.g., acute pulmonary embolism)
- Head trauma or medications that can cause dysfunction of respiratory drive
- Neuromuscular disease (not related to spinal cord injury [SCI])
- Patients with SCI with lesions above C3 present with diaphragmatic failure
  Note: Chronic respiratory failure—considered when ventilatory failure exceeds 30 days

Pulmonary Function Testing

- The magnitude of functional impairment can be measured using PFT.
- Respiratory excursions during normal breathing as well as during maximal inspiration and expiration are observed.
- Evaluation of lung volume changes can be used to classify respiratory dysfunction into obstructive and restrictive pulmonary disease.

Lung Volume Definitions (Figure 9–2)

- **Vital capacity (VC):** The greatest volume of air that can be exhaled from the lungs after maximum inspiration
- **Forced vital capacity (FVC):** VC measured with the subject exhaling as rapidly as possible
- **Total lung capacity (TLC):** Amount of gas within the lungs at the end of maximal inspiration
- **Tidal volume (TV):** Amount of gas moved in normal resting inspiratory effort

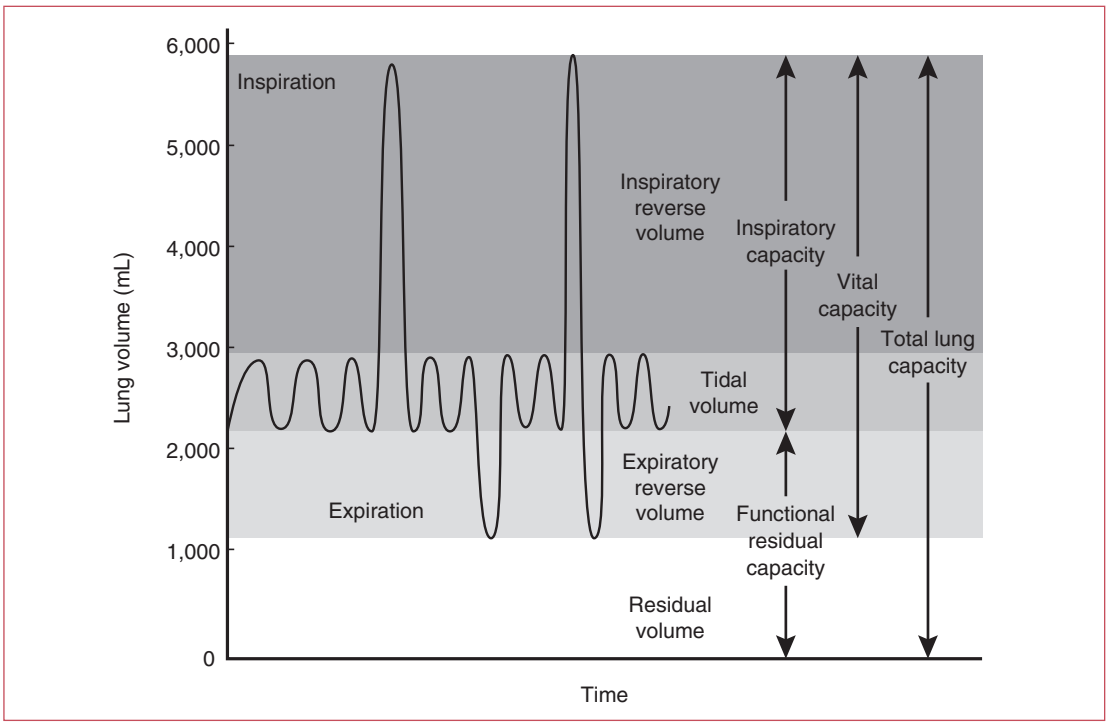


FIGURE 9–2 Illustration of respiratory excursions during normal breathing, maximal inspiration, and maximal expiration.

- **Functional residual capacity (FRC):** Amount of gas in the lungs at the end of normal expiration
- **Residual volume (RV):** Amount of gas in the lungs at the end of maximal expiration
- **Forced expiratory volume in 1 second (FEV_1):** Amount of air expelled in the first second of FVC
- **Maximal mid-expiratory flow rate (MMEF):** Average flow rate, between 25% and 75% of FVC
- **Maximal voluntary ventilation (MVV):** The maximum volume of air exhaled in a 12-second period in liters per second
- **Maximal static inspiratory pressure (PImax):** Static pressure measured near RV after maximal expiration
- **Maximal static expiratory pressure (PEmax):** Static pressure measured near TLC after maximal inspiration
- **Minute volume:** Volume of gas inhaled or exhaled per minute

🕮 Maximal Oxygen Consumption

- Expired gases during maximal exercise are collected and analyzed for oxygen content.
- $\dot{V}O_2$max: The maximal rate of oxygen consumption attainable during physical exertion.
- $\dot{V}O_2$max is expressed as a relative rate in milliliters (mL) of oxygen per kilogram (kg) of body mass per minute (mL/kg/min).
- Individual $\dot{V}O_2$max is dependent on body weight, age (peak is reached at approximately 20 years of age), sex (values for females are approximately 70% those of males), pulmonary function, cardiac output (CO), and inherent genetics (the most important).
- Training or the presence of pathologic conditions can affect this potential.
- Endurance exercise training increases $\dot{V}O_2$max, CO, and physical work capacity of untrained healthy individuals.

Can be calculated using the Fick equation:

$$\dot{V}O_2\text{max} = (\text{HR} \times \text{SV}) \times AVO_2 \text{ difference}$$

AVO_2, arterial venous oxygen; HR, heart rate; SV, stroke volume.

🕮 CLASSIFICATION OF RESPIRATORY DYSFUNCTION

- Based on PFTs, respiratory dysfunction can be classified as obstructive or restrictive.

Chronic Obstructive Pulmonary Disease

- Airflow obstruction in COPD is caused by a mixture of small airway disease (increased airway resistance and obstruction) and parenchymal destruction (decreased elastic recoil in the lung parenchyma). Chronic inflammation in the lung causes these structural changes and decreases elastic recoil (GOLD Guidelines, 2024).
- 🕮 Characterized by increased airway resistance due to bronchospasm, which may result in air trapping
- 🕮 Structural abnormalities in airways, alveoli, and pulmonary circulation cause ventilation/perfusion mismatching, resulting in different degrees of hypoxemia with or without hypercapnia (GOLD Guidelines, 2024).
- Hypercapnia occurs in progressive disease and is associated with loss or blunting of the hypoxic drive or acute decompensation.
- 🕮 Can be clinically associated with increased airway resistance, impaired expiratory airflow, and respiratory muscle fatigue. Flattening of the diaphragm seen on chest x-ray due to increased total and residual lung volumes.
- **Prevalence:** Approximately 6.4% of all Americans, but data vary widely due to differences in survey methods, diagnostic criteria, and analytical approaches (GOLD Guidelines, 2024).

- Chronic lower respiratory disease, primarily COPD, was the fourth leading cause of death in the United States in 2017 (Murphy et al., 2019).
- 50% have limitations in activity level, and 25% are limited to bed activities.

CAUSES OF CHRONIC OBSTRUCTIVE PULMONARY DISEASE

- Caused by a combination of factors:
  - Genetic predisposition
  - Respiratory infections
  - Tobacco: Cigarette/pipe, cigar, and marijuana smoking
  - Passive exposure to smoke
  - Biomass exposure
  - Air pollution and household air pollution
  - Occupational exposure to dust, chemicals, and fumes
- Cigarette smoke is the most common cause of COPD in high-income countries.
  - Causes chronic inflammation and decreased mucociliary clearance
  - Smokers are more likely to die from COPD than nonsmokers (3.5–25 times more likely).
  - Smoking cessation is linked to improvement in the following:
    - Improvement in symptoms
    - Variable improvement in pulmonary function, although there is a decline in the rate of loss of lung volume to a rate similar to nonsmokers
    - Decreased risk of respiratory tract infection
    - Decreased reduction in rate of loss of FEV_1 (long term)

TYPES OF CHRONIC OBSTRUCTIVE PULMONARY DISEASE

- Remember: All forms of COPD can involve air trapping (airflow obstruction).
- **Chronic bronchitis:**
  - Chronic mucus hypersecretion and respiratory infections as a result of tracheobronchial mucous gland enlargement
- Chronic cough and sputum for at least 3 months for 2 consecutive years in the absence of other conditions to explain symptoms (Agustí et al., 2023)
- **Emphysema (Figure 9–3):**

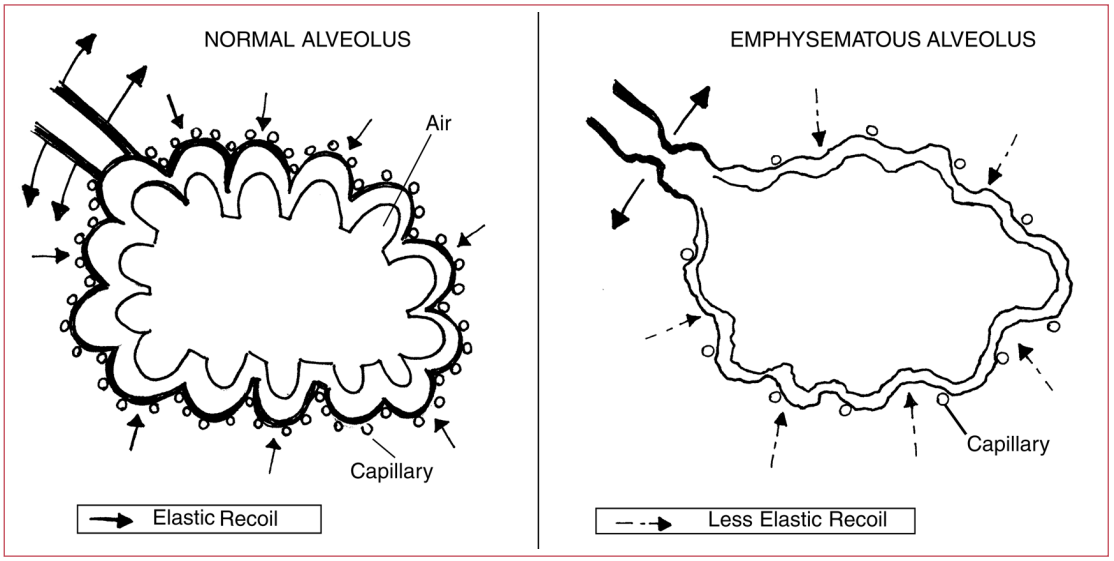


FIGURE 9–3 Normal single alveolus as compared with emphysematous single alveolus.

  - Distention of air spaces distal to the terminal nonrespiratory bronchioles with destruction of alveolar walls. This is secondary to the unimpeded action of neutrophil-derived elastase.
  - Destruction of the alveolar wall elasticity results in loss of lung recoil, leading to excessive airway collapse on exhalation and chronic airflow obstruction.
  - Decreased gas exchange surface area of the lung (i.e., alveolar membranes in association with $\dot{V}/\dot{Q}$ mismatch causes hypoxemia)
  - Chronic increase in pulmonary vascular resistance in the presence of pulmonary tissue hypoxia can lead to severe pulmonary artery hypertension (PAH) and right ventricular heart failure.
  - Oxygen is the only proven therapy that improves mortality in hypoxemic patients.
- 🕮 **Cystic fibrosis (CF):**
  - Autosomal recessive disease involving the chloride ion channels found in exocrine glands. Respiratory involvement is caused by failure to adequately remove secretions from the bronchioles, resulting in widespread bronchiolar obstruction and subsequent bronchiectasis, overinflation, and infection.
  - 🕮 Aerobic exercise for CF patients helps to increase sputum expectoration. Patients have increased ciliary beat with improved mucous transport.
  - Aerobic exercise also improves exercise capacity and respiratory muscle endurance, and reduces airway resistance by facilitating expectoration of retained secretions.
  - Chest physiotherapy can help to mobilize airway secretions.
  - 🕮 Exercise recommendations:
    - Mild to moderate CF lung disease: 70% to 85% maximal heart rate (HRmax), 30 to 45 min/day, three to five times per week
    - Severe CF lung disease: 60% to 80% HRmax, 20 to 30 min/day, five times per week
    - Add supplemental oxygen if saturation falls below 90% during exercise.
  - Possible risks associated with exercise and training include dehydration, hypoxemia, bronchoconstriction, pneumothorax, hypoglycemia, hemoptysis, esophageal bleedings, cardiac arrhythmias, rupture of liver and spleen, and spontaneous fractures.
- **Asthma:**
  - Hypertrophy of bronchial muscle, mucosal edema, and infiltration with eosinophils and mononuclear cells, which cause changes in the basement membrane. Patients with asthma are more likely to develop bronchitis. Asthma might be a risk factor for the development of COPD.
  - Episodic widespread narrowing of airways and paroxysmal expiratory dyspnea at night can occur.
  - 🕮 Evidence has shown that an inpatient pulmonary rehab program can significantly improve the degree of asthma control up to 3 months after the end of the rehabilitation program with additional benefits in symptoms (e.g., shortness of breath, coughing, sputum production), quality of life, psychological stress, and subjective ability to work.

🕮 GENERAL GUIDELINES TO ASSESSING FUNCTIONAL LIMITATIONS OF CHRONIC OBSTRUCTIVE PULMONARY DISEASE PATIENTS BASED ON PULMONARY FUNCTION TEST FINDINGS

- The magnitude of functional impairment in COPD patients can be assessed using PFTs.
- When the predicted FEV_1 is close to 4 L, the patient *should not* have a history of significant exercise impairment.
- Impairment usually develops when FEV_1 falls below 3 L/sec, but is variable.
  - FEV_1 between 2 and 3 L: The patient may experience mild exercise limitation (able to walk significant distances but not at high speed).
  - FEV_1 between 1 and 2 L: The patient may experience moderate degree of exercise impairment (intermittent rest periods are required to walk significant distances or to climb stairs).
  - FEV_1 <1 L: Severe exercise impairment (very short-distance ambulation)

Restrictive Pulmonary Disease

- 🕮 Impaired lung ventilation due to loss of normal elastic recoil of the lungs or chest wall. This can be associated with respiratory muscle dysfunction or stiffness of the chest wall or lung tissue itself, thereby resulting in increased work to breathe.
- Can be associated with variable levels of hypercapnia or hypoxia
- Almost all lung volumes are decreased, and flow rates are increased, owing to the loss of normal compliance.

🕮 CAUSES OF RESTRICTIVE PULMONARY DISEASE

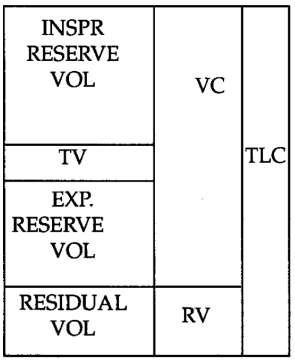
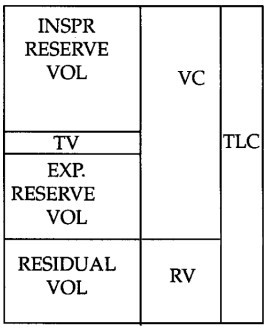
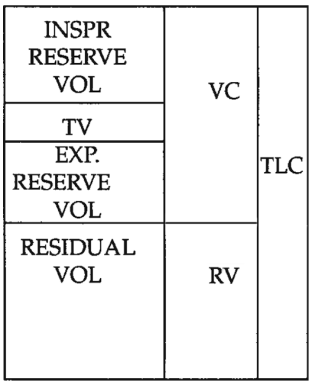
- **Intrinsic lung diseases (increased stiffness of lung tissue):**
  - Note: Intrinsic lung disease can lead to pHTN, right ventricular hypertrophy, and cor pulmonale.
  - Examples: Asbestosis, sarcoidosis, silicosis, and idiopathic pulmonary fibrosis
- **Extrinsic lung diseases (e.g., increased stiffness of chest wall):**
- Neuromuscular diseases:
  - Examples: Duchenne muscular dystrophy (DMD), amyotrophic lateral sclerosis (ALS), Guillain–Barré syndrome (GBS), myasthenia gravis (MG)
  - Weakness of respiratory muscles impairs the bellows activity of the chest wall, limiting ventilatory capacity and causing hypoventilation.
  - Respiratory muscle weakness causes impaired cough.
- Thoracic deformities (e.g., kyphoscoliosis):
  - If scoliotic angle is >90 degrees, patients have dyspnea.
  - If scoliotic angle is >120 degrees, patients present with hypoventilation and may have cor pulmonale.
- Pleural disease
- Ankylosing spondylitis (AS)—limited expansion of the chest wall
- Cervical SCI
- Obesity
- Surgical removal of lung tissue

Types of Extrinsic Restrictive Lung Disease

- **DMD (X-linked recessive inheritance):**
  - 🕮 Patients present with several respiratory complications including:
    - Atelectasis secondary to hypoventilation
    - Pneumonia
- **Poliomyelitis:**
  - A viral disease
  - Patients have a higher risk of restrictive lung disease and hypoventilation versus healthy individuals.
  - More likely to develop respiratory muscle weakness and skeletal deformities (scoliosis)
- **Chronic alveolar hypoventilation (CAH) with hypoxemia:**
  - Diseases that lead to respiratory muscle weakness, such as DMD, lead to hypoventilation in which not enough air enters the lungs. CAH is characterized by reduced arterial oxygen tension (PaO_2) and increased carbon dioxide tension ($PaCO_2$), leading to daytime fatigue, hypersomnia, and changes in psychological function.
  - A systematic review by Cochrane suggests that in people with chronic hypoventilation related to neuromuscular or chest wall diseases, nocturnal mechanical ventilation improves hypoventilation-related clinical symptoms, nocturnal mean oxygen saturation, daytime hypercapnia, and quality of life, and reduces the risk of unplanned hospitalization and mortality, compared with no ventilation.

  - Note: Noninvasive positive-pressure ventilation (such as bilevel positive airway pressure [BiPAP] Trilogy, and Life2000), or invasive ventilator-driven support via tracheostomy, can be utilized.
- **Ventilatory failure:**
  - Progressive weakness of respiratory musculature causes ventilatory limitation and impaired cough.
  - DMD patients also develop progressive scoliosis, which limits expansion of the chest wall and interferes with respiration.
- **Amyotrophic lateral sclerosis (ALS):**
  - Most common form of motor neuron disease that causes respiratory failure. Respiratory failure usually develops late in the disease and is the most common cause of death.
  - Respiratory muscle weakness causes restrictive ventilation and impaired cough.
  - If symptoms begin with limb weakness, the disorder may progress to respiratory failure in 2 to 5 years.
  - The American Academy of Neurology recommends measuring FVC at diagnosis and every 3 months thereafter to monitor disease progression.
  - FVC below 50% of predicted suggests the risk of imminent respiratory failure and the need for ventilatory support.
  - FVC may be falsely low with bulbar weakness due to inability to form a tight mouth seal during spirometry. Conversely, due to preserved lung volume, FVC may remain normal until there is substantial muscle weakness.
  - Further testing can be done by measuring transdiaphragmatic pressure (Pdi), which is considered to be the gold standard measure of diaphragmatic strength. It is obtained by measuring gastric and esophageal pressures with gastric and esophageal balloon catheters. Pdi is gastric pressure minus esophageal pressure.
- **Spinal deformities (kyphoscoliosis):**
  - Severe kyphoscoliosis limits expansion of the chest wall, reduces lung volumes, and compromises respiratory muscle efficiency.
  - If scoliotic angle is >90 degrees, patients often complain of dyspnea.
  - If scoliotic angle is >120 degrees, patients may develop overt hypoventilation and cor pulmonale.
- **Ankylosing spondylitis (AS):**
  - There is physical limitation to chest wall expansion secondary to the ankylosing process.
- **Cervical SCI:**
  - In SCI, respiratory dysfunction is related to three factors: Reduced VC (weak respiratory muscles, atelectasis), retention of secretions (increased secretions, ineffective cough), and autonomic dysfunction (increased secretions, bronchospasms, pulmonary edema).
  - Diaphragm is innervated by the phrenic nerve (C3–C5).
  - Spinal cord trauma sparing phrenic nerve innervation leaves diaphragm function intact and allows for spontaneous ventilation.
  - Complete lesions above C2 result in loss of function of diaphragm and intercostal muscles.
  - Lesions above C3 eliminate all but accessory muscles of breathing.
  - RV increases in C-spine injury.
  - These patients have difficulty clearing secretions and ventilatory failure can ensue.
  - Inspiratory muscle training (IMT) uses devices with spring-loaded valves that allow for expiration and offer resistance to inspiration. Weights placed on the abdomen or incentive spirometers can also be used to offer resistance to inspiration.
  - Although lower cervical and high thoracic cord lesions leave diaphragm function intact, they decrease intercostal and abdominal muscle function, which impairs expiratory muscle function. Weak expiration leads to impaired cough and secretion retention.
  - Expiratory muscle function can be improved by (a) strengthening the pectoralis major muscle (C5–C7 innervation) or (b) electrical surface stimulation to the abdominal muscles or lower thoracic medulla.

PULMONARY FUNCTION TESTING

NORMAL LUNG VOLUMES	RESTRICTIVE LUNG DISEASE	OBSTRUCTIVE LUNG DISEASE
		
Normal changes noted with aging.	**Key point: All volumes are decreased.**	**Key point: Air trapping occurs.**
• **Decreases in** VC MVV FEV_1 PO_2 • **FEV_1 decreased at a rate of 30 mL/year** • **No changes in** TLC PCO_2 • **Increases in** RV FRC	• **Increased stiffness of chest wall** Ankylosing spondylitis Cervical SCI Neuromuscular disease including: DMD, ALS, MG, GBS Kyphoscoliosis • **Increased stiffness of lung** Pulmonary edema Interstitial lung disease • **Increased elastic work of breathing** • **Decreases in** VC TLC RV[a] FRC FVC MVV (decreases in severity) All volumes are decreased; this is distinctive for restrictive lung disease • **FEV_1 is normal.**	• **Limitation in expiration before air is fully expired** COPD Cystic fibrosis Asthma Chronic bronchitis • **Flattening of the diaphragm** Increase in airway resistance Expiratory effort Respiratory muscle fatigue Impaired gas exchange as a result of air trapping leads to respiratory muscle fatigue • **Decreases in** VC FEV_1 MVV FVC FEV_1 decreases 45–75 mL/year in COPD patients • **Increases in** RV FRC TLC (but TLC can also be normal)

Note: MVV decreases in most pathologic states and aging.
Key: Refer to section titled Lung Volume Definitions earlier in this chapter.
[a]RV increases in cervical SCI.
ALS, amyotrophic lateral sclerosis; COPD, chronic obstructive pulmonary disease; DMD, Duchenne muscular dystrophy; FEV_1, forced expiratory volume in 1 second; FRC, functional residual capacity; FVC, forced vital capacity; GBS, Guillain–Barré syndrome; MG, myasthenia gravis; MVV, maximal voluntary ventilation; RV, residual volume; SCI, spinal cord injury; TLC, total lung capacity; TV, tidal volume; VC, vital capacity.

LUNG VOLUME CHANGES IN UNIQUE MEDICAL CONDITIONS

Tobacco Use Versus Normal Aging

- Normal rate of decrease in FEV_1 is approximately 30 mL/year.
- In smokers, this can increase to two to three times this value.

- Smokers <35 years old can increase lung function if they quit smoking.
- If a patient is >35 years old and quits smoking, the rate of decline of lung function slows to the normal rate associated with aging, and some improvement in function can occur.

Cervical Spinal Cord Injury

- Cervical SCI patients have restrictive lung disease patterns.
- 🕮 Pulmonary changes seen in C5 tetraplegics:
  - Diaphragm remains intact and the expiratory muscles are paralyzed.
  - Patients retain approximately 60% of their inspiratory capacity and ventilate well, but have a weak cough and difficulty clearing secretions during respiratory infections.
  - All volumes are greatly reduced because of limited expansion of the chest wall.
  - Decreased TLC and VC
  - Increased RV
- 🕮 In SCI patients, the abdominal contents may sag due to the greater strength of the diaphragm relative to the weakness of the abdominal wall muscles. This decreases diaphragmatic excursion and the VC in the sitting position.
- The reduction in VC is most severe in tetraplegics with cervical cord injury and during the acute injury period. Severity of reduction increases with the higher level of injury. A study by Maloney (1979) reported that in the sitting position the use of an abdominal binder improved VC (**Figure 9–4**).
- The goal of pulmonary rehabilitation of the SCI patient is to:
  - Increase VC
  - Maintain good pulmonary hygiene
  - Optimize secretion mobilization and management
  - Manage and treat any detected dysphagia
  - Subjectively improve dyspnea as it relates to patients' functional mobility and self-care
  - Reduce average number of hospital stays

Duchenne Muscular Dystrophy

- VC plateaus between 1,100 and 2,800 mL between 10 and 15 years of age.
- Independent of chest deformity, the VC is then lost at a rate of 200 to 250 mL/year. The rate of loss tapers below 400 mL.
- 🕮 No clear guidelines have been established for determining the point at which ventilatory support should be instituted in patients with DMD, but various studies suggest the following:
  - Dyspnea at rest
  - 45% predicted VC
  - Maximal inspiratory pressure <30% predicted
  - Hypercapnia

Amyotrophic Lateral Sclerosis

- Routine PFTs, especially functional VC, should be monitored closely in ALS.
- The earliest changes noted are decreases in maximum inspiratory and expiratory muscle pressures, followed by reduced VC and maximum breathing capacity.

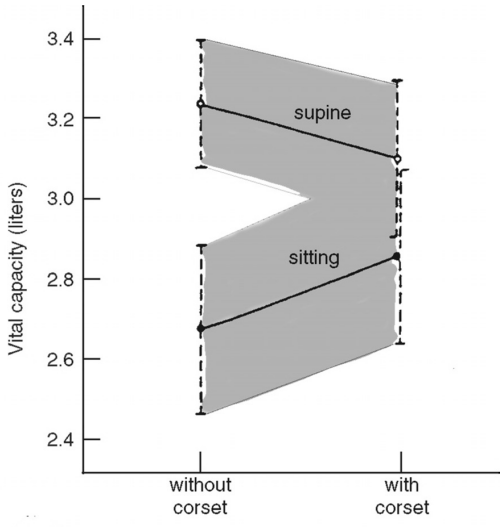


FIGURE 9–4 Comparison chart of actual vital capacities with and without wearing a corset in supine and sitting position. The chart shows the interaction of the position and the use of the corset. Note the significant difference in vital capacity in the patient without the corset in the supine versus the seated position. The vital capacity is improved in the seated position with corset.

- When VC falls to 25 mL/kg, the ability to cough is impaired, which increases the risk of aspiration pneumonia.
- FVC is the best prognostic indicator for noninvasive ventilation in patients with ALS. Patients may lose VC at a rate of 1,000 mL or more per year.
- Various changes in blood gases including hypoxemia, and hypercarbia to a variable extent, can be seen. Hypercapnia may occur as disease progresses and as VC decreases (Bach, 1996).

REHABILITATION MANAGEMENT OF THE CHRONIC OBSTRUCTIVE PULMONARY DISEASE PATIENT

1. Nutritional Status Evaluation

- Respiratory muscle weakness is often associated with metabolic deficits.
- Decreased magnesium, calcium, and potassium and hypophosphatemia are associated with respiratory muscle weakness, which is reversible after electrolyte replacement.
- Serum albumin level correlates with severity and chronicity of disease, degree of hypoxemia, and spirometric values. It indicates visceral protein depletion and is a good predictor of rehabilitation potential.
- Protein supplementation ≥1.7 g/kg of body weight per day is associated with an increase in nitrogen retention and physiologic improvement.
- Impaired nutritional status is associated with increased morbidity and mortality.
  - More frequent infections due to impaired cell-mediated immunity
  - Decreased macrophage action in the pulmonary alveolar region
  - Increased bacterial adherence and colonization in upper and lower airways
  - *Pseudomonas* species commonly colonize in patients with poor nutrition in a nosocomial environment.
- Poor nutritional state affects lung repair mechanisms, including surfactant synthesis.
- Poor nutrition can also lead to generalized weakness, affecting respiratory function, and ultimately hypercapnic respiratory failure as well as problems with weaning from mechanical ventilation.
- Impaired nutritional status is associated with increased hospitalizations, poor exercise tolerance, and worsened quality of life (GOLD Guidelines, 2024).
- COPD patients are encouraged to increase fluid intake.
- Also evaluate for obesity. This condition increases the work of the respiratory system, particularly during weight-bearing activities. Encourage weight reduction.

2. Pharmacologic Optimization Prior to Rehabilitation Program

- 🕮 For dyspnea and to decrease exacerbations of COPD, we can follow the 2024 GOLD Guidelines for management recommendations:
  - Inhaled anticholinergics (e.g., ipratropium [Atrovent®], umeclidinium bromide [Incruse®], and tiotropium [Spiriva®]) block muscarinic receptors.
  - Short-acting inhaled beta-2 agonists for emergency use
  - Note: Anticholinergics, for example, ipratropium, can be used alone or added to a regimen including beta-2 agonists. They block smooth muscle muscarinic receptors.
- Inhaled steroids can decrease the frequency of COPD exacerbations in certain COPD patients and are also used for asthma exacerbations. It is important to instruct the patient on how to administer it since >60% use inhalers incorrectly.
- In patients with COPD not receiving inhaled corticosteroids, regular treatment with mucolytics such as *N*-acetylcysteine (NAC) may reduce exacerbations and modestly improve health status (GOLD Guidelines, 2024). May also use expectorants for secretion control.
- Theophylline has a bronchodilator effect. Enhanced inspiratory muscle function has been reported in patients treated with methylxanthines, but whether this reflects a reduction in gas trapping or a primary effect on the respiratory skeletal muscles is not clear. All studies that have shown efficacy of theophylline in COPD were performed with sustained-release preparations. There is evidence for a modest bronchodilator effect compared with placebo in stable COPD (GOLD Guidelines, 2024). Theophylline is no longer considered first- or second-line therapy

for asthma or COPD. Use is limited by potential toxicity (Global Strategy for the Diagnosis, Management, and Prevention of Chronic Obstructive Pulmonary Disease, 2007).
  - 🕮 Young patients with moderate asthma who have tried beta-2 agonists during exercise as well as mast cell stabilizers or leukotriene inhibitors may benefit from theophylline use for exercise-induced asthma/bronchospasm.
- **Exercise-induced bronchospasm (EIB)**:
  - Transient increase in airway resistance that may appear 5 to 15 minutes after initiation of exercise. Caused by loss of heat, water, or both from the lungs during exercise as the result of hyperventilation.
  - Symptoms: Cough, wheezing, chest tightness, or pain (upset stomach or sore throat) *after* the first 5 minutes of exercise
  - Management of EIB:
    - Short-acting bronchodilators (beta-2 agonists) with the use of anti-inflammatory agents (inhaled corticosteroids) in the event of frequent recurrences, taken 15 to 30 minutes prior to starting exercise
  - Inhaled corticosteroids, with or without a bronchodilator, are the next step in treatment when a suboptimal response and abnormal PFTs are obtained. Use for weeks to obtain maximal effect.
  - Anticholinergics become a tertiary agent in EIB.
  - Nonpharmacologic management of EIB includes:
    - Increasing physical conditioning
    - Warm-up period for at least 10 minutes prior to exercise
    - Cover the mouth and throat during cold weather.
    - Exercise should be performed in a humidified environment, when possible.
    - Avoid pollutants and aeroallergens.
    - Lower the intensity of exercise; cool down prior to stopping exercise.
    - Do not exercise for at least 2 hours following a meal.

3. Supplemental Oxygen Use

- Low-flow nasal supplemental O_2 can be used during therapy to reduce dyspnea and improve exercise performance, especially in patients with documented coronary artery disease (CAD).
  - 🕮 O_2 is recommended for patients who desaturate during exercise. The most accepted guideline for O_2 use during exercise is if the patient exhibits an exercise-induced SaO_2 below 88% (CMS.gov, 2023).
  - Benefit of O_2 in patients without resting hypoxia is questionable.
- Inspiratory phase or pulsed oxygen therapy, especially if provided transtracheally, decreases mucosal drying and discomfort. O_2 delivery is of 0.25 to 0.4 L/min compared with 2 to 4 L/min via face mask or nasal cannula (rarely employed clinically).
- Supplemental O_2 use is also recommended for patients with a continuous PO_2 of 55 to 60 mmHg.
- The GOLD Guidelines (2024) recommends long-term oxygen therapy for stable patients who have:
  - $PaO_2 \leq 55$ mmHg or $SaO_2 \leq 88\%$ with or without hypercapnia confirmed twice over a 3-week period, or
  - PaO_2 of 55 mmHg or SaO_2 of 88% if there is evidence of pHTN, peripheral edema, or suggestive of congestive heart failure (CHF) or polycythemia with hematocrit >55%
- 🕮 Benefits of home oxygen use include:
  - Reduction in polycythemia
  - Improvement in pHTN
  - Reduction of the perceived effort during exercise
  - Increased survival
  - Improvement in cognitive function
  - Reduction in hospital needs
  - Decreased blood pressure and pulse in patients with COPD, who have increased sympathetic activity and reduced baroreflex sensitivity
- Cessation of smoking should be mandated.

- Patients with mild to moderate daytime hypoxemia often have significant nocturnal desaturation. Home overnight oximetry can be used to diagnose nocturnal oxyhemoglobin desaturation and assist in oxygen prescription; guidelines for sleep supplemental oxygen have not yet been established, but the general consensus is that nocturnal oxygenation should remain about 88% or home oxygen is necessary.
  - Consider a sleep study.
- Noninvasive ventilation may be used in patients with elevated CO_2 levels.

4. Training in Controlled Breathing Techniques

- COPD patients exhibit an altered pattern of respiratory muscle use. The rib cage inspiratory muscles generate more pressure than the diaphragm. Expiratory muscles are also involved.
- 🕮 Controlled breathing techniques are used to reduce dyspnea, reduce the work of breathing, and improve respiratory muscle function and pulmonary function parameters. Different types may be used in patients with obstructive pulmonary disease and restrictive disease.

TECHNIQUES TO IMPROVE PULMONARY FUNCTION PARAMETERS

- ***Diaphragmatic breathing:***
  - Used to reverse altered pattern of respiratory muscle recruitment in COPD patients.
  - Patient uses the diaphragm and relaxes abdominal muscles during inspiration.
  - Lying down or at 15% to 25% head-down position, the patient places one hand over the thorax below the clavicle to stabilize the chest wall and the other over the abdomen.
  - The patient takes a deep breath and expands the abdomen using the diaphragm.
  - Feedback of abdominal and rib cage movement is obtained through hand placement as described previously.
  - 🕮 Benefits: Increased TV, decreased FRC, and increase in maximum oxygen uptake.
- ***Segmental breathing:***
  - Obstructions such as tumors and mucous plugs should be cleared prior to practicing this technique.
  - The patient is asked to inspire while the clinician applies pressure to the thoracic cage to resist respiratory excursion in a segment of the lung.
  - As the clinician feels the local expansion, the hand resistance is decreased to allow inhalation.
  - This facilitates expansion of adjacent regions in the thoracic cavity that may have decreased ventilation.

TECHNIQUES TO REDUCE DYSPNEA AND THE WORK OF BREATHING

- 🕮 ***Pursed-lip breathing:***
  - Patient inhales through the nose for a few seconds with the mouth closed, then exhales slowly for 4 to 6 seconds through pursed lips. Expiration lasts two to three times as long as inspiration.
  - By forming a wide, thin slit with the lips, the patient creates an obstruction to exhalation, slowing the velocity of exhalation and increasing mouth pressure.
  - 🕮 Benefits: Prevents air trapping due to small airway collapse during exhalation and promotes greater gas exchange in the alveoli. Increases TV and reduces dyspnea and the work of breathing in COPD patients. When added to diaphragmatic breathing, it reduces the RR and can improve arterial blood gases (ABGs; Bach, 1996).

5. Airway Secretion Management Program

- ***Controlled cough:***
  - The patient assumes an upright sitting position, inhales deeply, holds the breath for several seconds, contracts the abdominal muscles ("bears down," increasing intrathoracic pressure), then opens the glottis and rapidly and forcefully exhales while contracting the abdominal muscles and leaning slightly forward.
  - This is repeated two or three times and followed by normal breaths for several minutes before attempting controlled cough.
  - Coughing generates high expulsive forces promoting secretion retention and may exacerbate air trapping; also leads to fatigue if the cough is weak.

- *Huffing (huff coughing):*
  - An alternative is huffing—following a deep inhalation, the patient attempts short, frequent forceful exhalations by contracting the abdominal muscles and saying "ha, ha, ha."
  - The glottis remains open during huffing and does not increase intrathoracic pressure; helpful in COPD patients whose airways can collapse. This is a more efficient means of secretion removal.

Secretion Mobilization Techniques: Postural Drainage, Percussion, Vibration

- *Indications:*
  - Sputum production >30 mL/day
  - Aspiration
  - Atelectasis
  - Moderate sputum production in debilitated patients who are unable to raise their own secretions

POSTURAL DRAINAGE

- Utilizes gravity-assisted positioning to improve the flow of mucous secretion out of the airways
- The affected lung segment is placed at the *highest position* relative to the rest of the lung to optimize oxygenation and drainage.
- Best done after awakening in the morning (secretions accumulate at night) and 1 to 2 hours after meals to avoid gastroesophageal reflux

POSITIONS FOR POSTURAL DRAINAGE (FIGURE 9–5)

- A commonly used position is the Trendelenburg position (feet higher than the head), which can be done with the patient lying supine or prone and with different postural variations, such as side-lying or trunk bending.

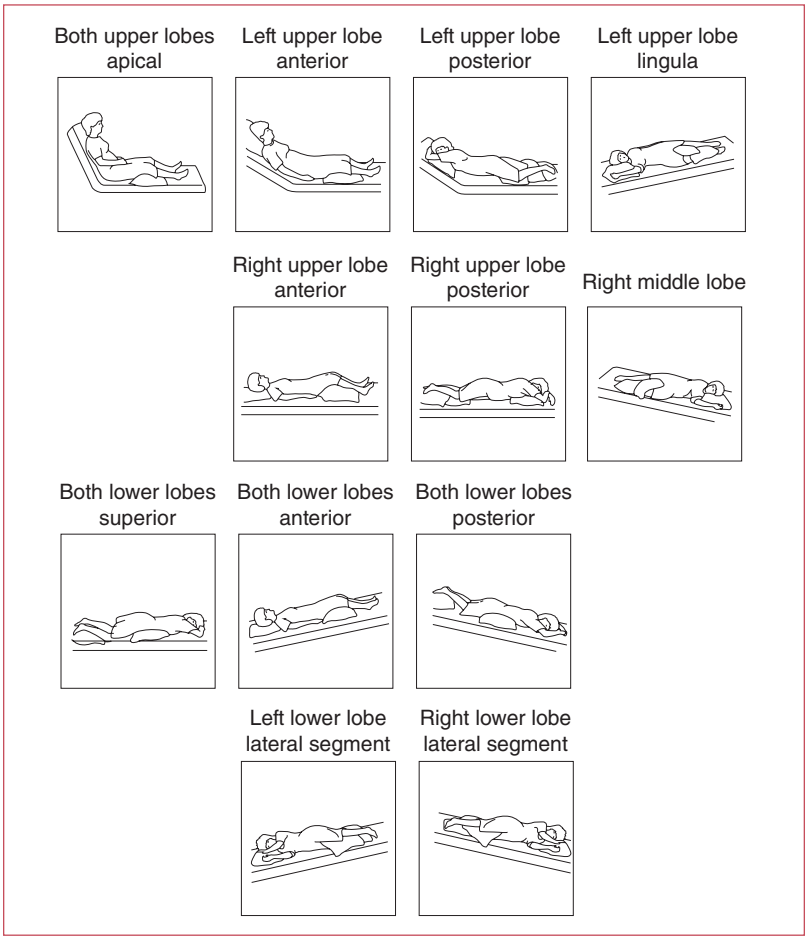


FIGURE 9–5 Postural drainage positions.

- *To drain the upper lobes:*
  - Patient is positioned sitting up.
  - Exceptions:
    - Right anterior segment—patient supine
    - Lingular—patient in lateral decubital Trendelenburg
    - Both posterior segments—prone
- *To drain the right middle lobe and lower lobes:*
  - Patient is positioned in the lateral decubital Trendelenburg.
  - Exceptions:
    - Superior segment of the lower lobe—patient prone with buttocks elevated
    - Posterior lower segment—patient in prone Trendelenburg position with buttocks elevated
    - Anterior segment—supine Trendelenburg
- Precautions for postural drainage:
  - Trendelenburg positioning (head-down tilt) can range from 10 to 45 degrees. COPD patients can tolerate up to a 25-degree tilt.
- Avoid in:
  - Pulmonary edema
  - CHF
  - Hypertension (HTN)
  - Dyspnea
  - Abdominal problems—hiatal hernia, obesity, recent food ingestion, abdominal distention
- Side-lying position contraindications:
  - Axillofemoral bypass graft
  - Musculoskeletal pain—for example, rib fractures

🕮 *POSTURAL CHANGES*

- Postural changes not only help with secretion mobilization but also affect the work of breathing by changing the mechanical load on the respiratory muscles and the oxygen supply and consumption in these areas.

1. *Mechanical load*—Pressure changes related to position:
   - Upright position—abdominal contents remain in low position due to gravity; diaphragm can compress them easily.
   - Supine position—redistributes abdominal contents. Diaphragm is in a slightly longer resting position further up into the thorax.
   - Trendelenburg—diaphragm at its longer resting position, displaced by the weight of the abdominal contents into the thorax.
     - With progression from the sitting to a Trendelenburg position, the diaphragmatic work of breathing is increased due to the increased abdominal content load. The diaphragm will accommodate to the increase in load by increasing its contraction.
   - In obesity, the external load of the abdominal muscles may be greater than the muscle's capacity of contraction.
   - In neuromuscular disease, the muscles may not be able to generate tension against the abdominal content load, requiring changes in posture to assist in breathing. This is also valid for COPD patients where postural changes can affect the diaphragmatic mechanical response.
   - 🕮 The weight of the pulmonary tissue also contributes to overall pressure on the most dependent alveoli. The dependent alveoli expand in size when changing from sitting to supine position, increasing ventilation at the base of the lung.
2. *Blood flow*—gravity-dependent:
   - Maximum flow is greatest at the most gravity-dependent portions of the lung.
   - Upright sitting—$\dot{V}/\dot{Q}$ mismatch, most effective at the middle lung fields.
   - The lower lobes of the lung are preferentially perfused, while the upper lobes are preferentially ventilated. With inspiration, ventilation to the lower lobes increases.
   - In some patients, changing from supine to prone positioning displaces the weight of the abdominal contents, reversing blood flow distribution to the anterior segments.

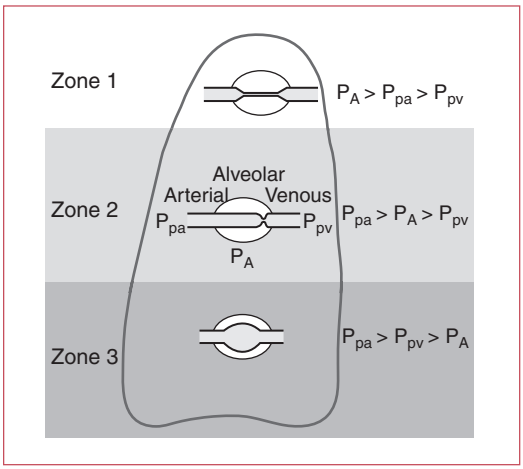


FIGURE 9–6 The three-zone model of the lung: The difference in blood flow distribution is based on the pressure affecting the capillaries.

Zone 1: Alveolar pressure (P_A) exceeds pulmonary artery pressure (P_{pa}), and no flow occurs because the vessels are collapsed.
Zone 2: Pulmonary arterial pressure (P_{pa}) exceeds alveolar pressure but alveolar pressure (P_A) exceeds pulmonary venous pressure (P_{pv}). The arterial–alveolar pressure difference ($P_{pa} - P_A$) determines the flow in Zone 2. This steadily increases down the zone.
Zone 3: Pulmonary venous pressure (P_{pv}) exceeds alveolar pressure and flow is determined by the arterial venous pressure (P_{pa}) difference ($P_{pa} - P_{pv}$), which is constant down this pulmonary zone. Note the pressure across the vessel walls increases down the zone so their caliber increases. As the caliber of the vessel wall increases, so does the flow.

- 🕮 The difference in blood flow distribution is based on the pressure affecting the capillaries (**Figure 9–6**).
- The pressure of the surrounding tissues can influence the resistance to blood flow through the capillaries.
- Blood flow depends on pulmonary artery pressure, alveolar pressure, and pulmonary venous pressure.
- The perfusion of the lung is dependent on posture.
- The perfusion of the three-zone model of the lung in the upright position is described as follows:
  - Zone 1: Ventilation occurs in excess of perfusion.
  - Zone 2: Perfusion and ventilation are fairly equal.
  - Zone 3: This is the most gravity-dependent region of the lung where pulmonary artery pressure > pulmonary venous pressure > alveolar pressure.
    - See **Figure 9–7A** for perfusion zones and the effect of posture on perfusion.

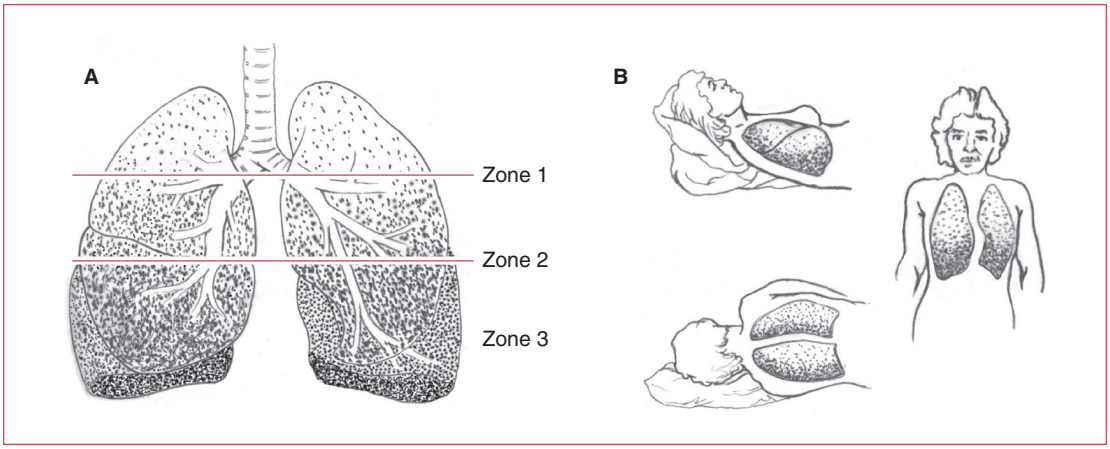


FIGURE 9–7 (A) Perfusion of the lung is dependent on posture. This diagram shows the perfusion of the lung in the upright position. (B) Perfusion of the lung is affected by positioning of the patient. The gravity-dependent segments have the greatest amount of perfusion.

- 🕮 When changing from a sitting to supine position, venous pressure increases in relation to the arterial pressure in dependent areas of the lung.
- Blood flow is governed by the pulmonary arterial to venous difference.
- When supine, the apical blood flow increases, but the bases remain virtually unchanged. There is an almost uniform blood flow throughout the lung. However, posterior segment flow will exceed anterior segment perfusion in this position.
- The normal ratio of ventilation/perfusion is 0.8. Areas of low ratios (perfusion > ventilation) act as a shunt. Areas of high ratios act as dead space.

PERCUSSION

- Mechanical percussor or a cupped hand can be used to rhythmically strike the thoracic cage during the entire respiratory cycle to loosen mucus within the lungs.
- Delivered at a frequency of 5 Hz for 1 to 5 minutes or longer over the chest area desired to be drained. Used on patients who are unable to mobilize and expectorate excess secretions or to help expand areas of atelectasis.
- *Precautions*:
  - Coagulation disorders
  - Anticoagulation therapy
  - Platelet count below 50,000
  - Fractured ribs
  - Flail chest
  - Severe osteoporosis
- *Contraindications:*
  - Cardiovascular instability or failure
  - Aortic aneurysm
  - Increased intracranial pressure
  - Increased intraocular pressure
  - Cannot do percussion over a tumor

VIBRATION

- Rapid shaking back and forth (not downward) on the thorax over a segment of the lung, causing mucus mobilization. Applied to the thorax or airway to facilitate secretion elimination.
- Can be applied manually or with a mechanical vibrator
- *Mechanical:*
  - Vibrator can be used at frequencies ranging from 10 to 15 Hz, up to 170 Hz.
  - Most animal studies favor the 10 to 15 Hz frequency range.
  - Uses very little or no pressure on the thorax and constitutes an alternative in cases where percussion is contraindicated.
  - The effects of mechanical chest percussion and vibration are frequency-dependent.
  - Side effects of percussion and vibration can include increased obstruction to airflow in COPD patients.

PREOPERATIVE AND POSTOPERATIVE CHEST THERAPY PROGRAM

- Airway clearance and secretion mobilization techniques can be applied prior to surgery and after the procedure.
- 🕮 A preoperative and postoperative chest therapy program has the following advantages:
  - Decreases the incidence of pneumonia
  - Reduces the probability of developing postoperative atelectasis following thoracic and abdominal surgery

PREOPERATIVE CHEST THERAPY PROGRAM

- The patient is taught standard postoperative treatment, including use of an incentive spirometer and various splinting techniques.

- *Deep breathing*—taught with the patient in the semi-Fowler position, in which the abdominal muscles are slack. This allows greater diaphragmatic excursion. Most important modality of postoperative pulmonary hygiene.
- *Rolling*—allows patient mobility and minimizes trunk movement
- *Coughing*—decreased cough effectiveness can be a result of anesthesia
- Two-stage cough, preceded by a deep diaphragmatic breath. First cough raises the secretions, and second cough facilitates expectoration. May use splinting techniques for coughing, splinting the surgical incision with the use of a pillow or hands.
- *Huffing*—see earlier
- *Incentive spirometry*—provides the patient with visual feedback of the air volume inspired during a deep breath. Patients practice deep inspiration every hour in addition to their chest physical therapy sessions.

POSTOPERATIVE CHEST THERAPY PROGRAM

- Most therapy programs start on postoperative day 1. Diaphragmatic and segmental breathing are used to assist the ventilator.
- Breathing exercises are provided.
- Secretion management techniques include postural drainage, vibration, and percussion.
- If the patient underwent abdominal surgery, one hand is placed between the incision site and the area to be percussed to decrease discomfort during the treatment. A pillow over the incision may also be used.
- Vibration is preferred postoperatively because it is less traumatic.
- These treatments are contraindicated in patients with cardiac or hemodynamic instability or in cases of pneumothorax.

6. Therapeutic Exercises

- Used to improve respiratory muscle endurance, strength, and efficiency

INSPIRATORY RESISTIVE LOADING

- Uses an inspiratory muscle trainer. The patient inhales through its inspiratory orifices, which progressively decrease in size. Exhalation is performed without resistance.
- Treatment is provided one to two times per day for approximately 15 to 30 minutes, with a rate of 10 to 20 breaths per minute. If the patient is able to tolerate 30-minute sessions, the intensity is increased by varying the orifice size. To increase endurance and orifice size, a longer exercise duration is chosen.

THRESHOLD INSPIRATORY MUSCLE TRAINING

- A threshold loading device allows inspiration only after a predetermined mouth pressure is reached. Produces inspiratory resistance without relying on inspiratory flow rates. Benefits include increased ventilatory strength and endurance.
- IMT has been proven beneficial in patients with CF, where FVC, TLC, and inspiratory muscle strength have been improved.
- IMT has appeared to prevent the weakness associated with steroid use in patients with this type of medication, as documented in one controlled study.
- In patients with asthma, a reduction in asthma symptoms has been noted in addition to the documented improvement in the inspiratory muscle strength and endurance. A reduction in hospitalizations and emergency room visits, increase in school and work attendance, and reduction in medication use have also been found.

7. Instruction on Reconditioning Exercises

- This type of exercise allows the patient to increase the ability to perform ADLs. The patient is engaged in a progressive program for which they are made responsible.

- Activities may include aerobic conditioning (bicycle, pool exercise program, walking, stair climbing, calisthenics), range of motion (ROM) exercises (coordinated with diaphragmatic breathing), and upper extremity strengthening exercises.
- A daily 12-minute walk with a record of time spent and distance achieved; 15 minutes per day of inspiratory training is also advised. The 12-minute walk can be used to estimate exercise tolerance.
- Pulse parameters include increase of at least 20% to 30% during the activity with a return to baseline within 5 to 10 minutes after exercise.
- The program is reevaluated weekly for 10 to 12 weeks, and modifications are made along with patient education.
- Upper extremity exercise reduces the metabolic demand and increased ventilation associated with arm elevation and resultant dyspnea.
- Unsupported upper extremity activities produce the most benefits, including decreased O_2 consumption. These types of activities include self-care, lifting, reaching, carrying, and athletic activities.
- All exercises should be performed to tolerance (symptom-limited, subjective dyspnea).
- Precautions:
  - Hold exercise for a heart rate (HR) >120 beats per minute (bpm).
  - Hold exercise if the patient has premature beats >6 bpm.
  - Hold exercise for oxygen saturation <92%. If the patient desaturates during exercise (<90%), the patient may use supplemental O_2 to enhance exercise performance. This also protects patients with CAD from dysrhythmia.
- 🕮 Aerobic exercise in patients with CF may include:
  - Exercises involving the trunk muscles, such as sit-ups
  - Swimming
  - Jogging
- Exercise recommendations for patients with CF include aerobic activities three to five times per week for 20 to 45 minutes at intensity levels of 60% to 85% of maximum HR depending on the severity of their lung disease.
- 🕮 Patients with CF who participate in a structured running program show significant improvements in exercise capacity, respiratory muscle endurance, and a reduction in airway resistance. In addition, studies in children with CF have found increased sputum expectoration and an improvement in lung function after several weeks of strenuous regular aerobic exercise.

8. Instruct About Energy Conservation Techniques

- These include paced breathing, body mechanics, advanced planning and prioritizing activities, and the use of assistive medical equipment.

9. Muscle Rest Periods Should Be Added to the Exercise Program

- Monitor hypercapnia as an indicator of the need for a rest period.
- Ventilatory assistance provides relief to tired respiratory muscles, decreasing their energy expenditure. Diaphragm rest can be achieved by assisting ventilation noninvasively with the use of body ventilators, mouthpiece, or nasal intermittent positive-pressure ventilation (IPPV) or tracheostomy IPPV.
- Although assisting ventilation can exacerbate air trapping in COPD patients, the benefits of resting respiratory muscles and decreasing oxygen consumption may outweigh this in importance.

IN PARTICULAR, TWO GROUPS OF CHRONIC OBSTRUCTIVE PULMONARY DISEASE PATIENTS MAY BENEFIT FROM VENTILATORY ASSISTANCE DURING REHABILITATION

1. Medically and psychologically stable patients who require assistance around the clock, usually by tracheostomy route

2. Patients with need of nocturnal assistance only. Nocturnal use of ventilators supports weak respiratory muscles.
   – Potential benefits include increased VC, respiratory muscle strength and endurance, and decreased need for hospitalizations.

VENTILATORY ASSISTANCE DEVICES FOR CHRONIC OBSTRUCTIVE PULMONARY DISEASE PATIENTS: POSITIVE VERSUS NEGATIVE AIRWAY PRESSURE BODY VENTILATORS

POSITIVE AIRWAY PRESSURE VENTILATION

- Can be IPPV, continuous positive airway pressure (CPAP), or BiPAP
- **IPPV** is the most common method of noninvasive support.
  – 🕮 Method of choice because the equipment is lightweight and portable; it is powered by either alternating current (AC) or direct current (DC); and no contact made with the individual other than the nasal–mouth interface or attachment to the tracheostomy tube
  – For mouthpiece IPPV, a mouthpiece is set up near the mouth where the patient can easily grab it up to six to eight times a minute for full ventilatory support. It is an ideal inspiratory muscle aid for daytime use in selected cases.
  – For nocturnal use, possibilities include nasal IPPV with CPAP mask (adequate seal is a problem) or mouthpiece IPPV with lip seal retention (seal is adequate, but patient cannot talk).
- 🕮 **CPAP** may be used to help maintain patent airways in patients with sleep disordered breathing (OSA), COPD, morbid obesity with hypoventilation, and hypoxic respiratory failure associated with CHF in which CPAP augments CO and improves $\dot{V}/\dot{Q}$ matching. It produces splinting of the pharyngeal airway with positive pressure delivered through a nose mask. CPAP provides higher oxygen stores, improves upper airway patency, and is one of the most cost-effective and least toxic forms of medical therapy. However, CPAP may decrease venous return and increase abdominal muscle effort. Additionally, long-term compliance with CPAP is low due to increased anxiety and disruptions in normal breathing patterns. Patients may also have mask interface-related skin changes (e.g., abrasions, pressure sores, contact dermatitis, etc.), aerophagia, sinus pain, oral and nasal dryness, and even tooth decay. It is also unavailable to individuals who are not spontaneously breathing.
- **BiPAP** permits independent adjustment of inspiratory (inspiratory positive airway pressure [IPAP]) and expiratory positive airway pressure (EPAP). Similarly to CPAP, this is used in patients with OSA and COPD, but can be utilized for individuals with poor respiratory drive. The variable pressure of BiPAP lowers pressure against which a patient exhales and therefore decreases both abdominal muscle recruitment and respiratory discomfort. The greater level of pressure assist also decreases inspiratory flow limitations in the upper airway. Compared with CPAP, BiPAP provides greater TV and unloading of the respiratory muscles. However, BiPAP has similar mask-interface-related issues and adherence difficulty as seen with CPAP.
- Positive-pressure ventilation via face mask may cause gastric distention and subsequently affect diaphragmatic function, compromising respiratory function.
- In patients with disorders of consciousness, concomitant noninvasive ventilation and enteral feedings via nasogastric tube (NGT) may increase risk of airway complications (e.g., vomiting, desaturation, mucus plug, aspiration pneumonia) and are associated with longer use of noninvasive ventilation.

NEGATIVE-PRESSURE BODY VENTILATORS → (RARELY USED TODAY)

- See also "Use of Noninvasive Ventilation" in the section that follows for further details.
- Used during the day or night, negative-pressure body ventilators (NPBV) have provided the following benefits:
  – Improved respiratory endurance with decrease in dyspnea
  – Improved quality of life, 12-minute walking distance
  – Improved Pdi and maximum inspiratory and expiratory pressures

- NPBV assists respiratory muscles by creating subatmospheric pressure around the thorax and abdomen.
- NPBV is also an alternative to intubation and tracheostomy for patients with acute respiratory failure (Bach, 1996).

REHABILITATION OF THE PATIENT WITH RESTRICTIVE LUNG DISEASE

- Respiratory complications are the most common cause of death in advanced restrictive lung disease.
- A major cause of acute respiratory failure for these patients is impaired secretion clearance.
- Rehabilitation of the patient with restrictive lung disease is based on prevention of complications and assistance with secretion management.

1. Patient Education

- Prevents development of pneumonia, respiratory failure, and subsequent intubation and mechanical ventilation
  - Importance of vaccinations should be stressed.
  - These include influenza, pneumococcal, and the possible use of antiviral agents.
- Avoid crowded areas or exposure to respiratory tract pathogens.
- Avoid sedatives at night and the risk of possible aspiration.
- Avoid excessive oxygen therapy. Central ventilatory drive can be suppressed, and the normal hypoxic pulmonary vasoconstriction can be inhibited, leading to a worsening $\dot{V}/\dot{Q}$ mismatch. These processes can lead to hypercapnia, and the risk of respiratory failure can be increased.
  - Studies indicate that excessive O_2 therapy can prolong hypopneas and apneas by 33% during rapid eye movement (REM) and by 19% otherwise, even in patients with mild neuromuscular disease.
- Avoid obesity and heavy meals.
- Develop goals and start planning for the future.

2. Maintenance of Adequate Nutritional State

- Respiratory muscle insufficiency can be exacerbated by hypokalemia.
- Patients with DMD have decreased total body potassium and commonly develop hypokalemia during acute illnesses.

3. Instruction in Controlled Breathing Techniques

GLOSSOPHARYNGEAL BREATHING

- This is a noninvasive method to support ventilation that can be used in the event of ventilator equipment failure.
- The patient takes a deep breath and uses the pistoning action of the tongue and pharyngeal muscles to project air boluses into the lungs. Rhythmic opening and closing of the vocal cords occurs with each air bolus.
- Each breath usually consists of six to nine air boluses (or up to 65), with each bolus consisting of 30 to 150 mL of air (usually 60–200 mL).
- Requires intact oropharyngeal muscle strength, and the patient should not be tracheostomized.
- 🕮 *Other uses of glossopharyngeal breathing:*
  - Enables the patient to breathe without mechanical ventilation (up to 4 or more hours if the lungs are normal; if lungs are affected, may only tolerate minutes). This time off the ventilator can be used to transfer to different types of aids.
  - Improves the volume of the voice and the rhythm of speech, allowing the patient to shout
  - Helps prevent microatelectasis
  - Allows the patient to take deeper breaths for more effective cough
  - Improves or maintains pulmonary compliance

DEEP BREATHING AND INSUFFLATIONS

- A program of air stacking hyperinflations two to four times per day with progressively increasing volumes helps prevent atelectasis and can benefit VC.
- Regular maximal insufflations can be provided with manual resuscitators, portable ventilators, and mechanical insufflators-exsufflators. A mouthpiece may be used, or a nosepiece may be provided, for larger volumes and when patients have weak oral muscles.

4. Secretion Management Techniques

MANUALLY ASSISTED COUGH

- The clinician's or the assisting person's heel of the hand or arm is placed at various sites along the anterior chest or abdomen to provide pressure and is coordinated with the patient's coughing or expiratory effort.
- Location of the areas of pressure:
  - Heimlich-type or abdominal thrust assist—patient in the side-lying position, pressure is applied at the navel while pushing up the diaphragm
  - Costophrenic assist—patient in any position, pressure is applied to the costophrenic angles
  - Anterior chest compression assist—patient lying on the side or the 3/4 supine position, pressure is applied to the upper and lower anterior chest
  - Counter rotation assist—pressure is applied to the pelvis or shoulder during inspiration followed by reversing the pressure direction to compress the thorax in all planes to facilitate expulsion

SUCTIONING

- Should be done in conjunction with other secretion clearing techniques, or when other techniques fail to remove secretions appropriately.
- May lead to complications such as airway membrane irritation and bleeding, airway edema and wheezing, hypoxemia, bradycardia or tachycardia, hyper- and hypotension, and increased intracranial pressure.
- ONLY suction as you withdraw the catheter.
- Chest percussion or postural drainage may also be used.

MECHANICAL INSUFFLATOR-EXSUFFLATOR

- Most effective method of mechanical assistance for secretion clearance in paralyzed patients
- A deep inspiration (positive-pressure insufflation) is provided via a mask or through the tracheal tube, followed by rapid controlled suction (negative-pressure exsufflation)
- Insufflation and exsufflation can be independently adjusted.
- A desired decrease in pressure from insufflation to exsufflation is approximately 80 cm H_2O. This may be sustained for 2 to 3 seconds. The duration of exsufflation is longer than with other methods of assistance.
- The decrease in pressure creates flows of approximately 7 to 11 L/sec, helping to bring secretions to the upper airways where those can be suctioned.
- It can be used in patients with scoliosis, dysphagia, impaired glottis function, and severe upper respiratory tract infections.
- Allows continued ventilatory support without tracheostomy and improves pulmonary volumes and SaO_2

5. Use of Noninvasive Ventilation

- Mechanically assisted ventilation provides respiratory muscle rest, decreasing the energy expenditure of the respiratory muscles.
- Body ventilation—includes positive, negative/positive, and negative-pressure ventilation.

POSITIVE-PRESSURE BODY VENTILATORS

- Provide positive pressure on the abdomen to assist diaphragmatic cephalad movement, promoting expiration. Passive inspiration occurs after removing the abdominal pressure.

- 🕮 *Intermittent abdominal pressure ventilator (IAPV):*
  - Examples: Pneumobelt (i.e., BachBelt), exsufflation belt
  - Abdominal corset containing a battery-operated rubber air sac. The air sac is cyclically inflated and deflated by a portable ventilator that delivers about 2.5 L of air at a time. When the air sac inflates, it compresses the abdomen, causing the diaphragm to move upwards and assist with expiration.
  - With cessation of airflow, the sac empties and the diaphragm descends due to gravity, causing inhalation. Inhalation with this device is mostly passive and dependent on gravity. Hence, it is only useful in the sitting or standing position. A trunk angle of 30 degrees or more from the horizontal is necessary for it to be effective, but 75 degrees is optimal.
  - Device is worn from the xiphoid to above the pelvic arch. Cycles are 40% inspiration and 60% expiration. Approximately 250 to 1,200 mL of TV can be provided.
  - This is the most useful mode of ventilation for wheelchair-bound patients with <1 hour of ventilator-free time during the day. Benefits also include liberating the mouth and hands for other activities.
  - Contraindicated in severe scoliosis and severe obesity. The patient should have a mobile abdomen.
  - Not useful in patients with decreased pulmonary compliance or increased airway resistance
  - Most beneficial when used during the day in addition to nocturnal noninvasive IPPV. Inspiration may be supplemented by the use of available inspiratory muscles and/or glossopharyngeal breathing.

NEGATIVE- AND POSITIVE-PRESSURE BODY VENTILATORS

- 🕮 ***Rocking bed:***
  - Rocks the patient along a vertical axis (15–30 degrees from the horizontal) utilizing the force of gravity to assist ventilation
  - When the head of the bed is up, inspiration is assisted by using gravity to pull the diaphragm down. This creates a negative pressure.
  - With the head down, exhalation assist is obtained. Cephalad movement of the abdominal contents pushes the diaphragm up with production of positive pressure.
  - It is used in patients with diaphragm paralysis with some accessory muscle function.
  - Benefits: Prevents venous stasis and improves clearance of bronchial secretions; weight shifting prevents the development of pressure ulcers and benefits bowel motility. Easy to apply.
  - Disadvantages: Heavy (not portable); not effective in patients with poor lung or chest wall compliance or in those with increased airway resistance.
- ***External oscillation ventilator (Hayek oscillator):***
  - Flexible chest enclosure (cuirass) with external oscillating ventilator
  - Pressure change is developed between the cuirass and the chest wall. Negative pressure helps chest wall expansion and inspiration. Positive pressure causes chest compression and aids expiration.
  - Inspiratory pressure is always negative, but the expiratory pressure can be adjusted to positive, zero or subatmospheric, and negative.
  - By increasing the number of oscillations per minute, it may be used for secretion clearance.
  - Patients with decreased lung compliance may use this type of assistance.

NEGATIVE-PRESSURE VENTILATORS

- Create intermittent extrathoracic pressure over the chest wall and abdomen, helping inspiration
- Main use is at night
- Provides rest to fatigued respiratory muscles
- Cor pulmonale may be prevented.
- The patient may be able to function during the day without respiratory assistance.

- Contraindicated in upper airway obstruction cases, where it may increase the frequency and severity of airway collapse and obstruction during the night. This may lead to obstructive apnea and desaturation.
- Not useful in children <3 years old due to recurrent pneumonias and atelectasis
- Not useful in patients with excessive airway secretions
- ***Tank ventilators (Emerson Iron Lung, Lifecare Porta-Lung):***
  - Patient's entire body is enclosed in a chamber that produces intermittent subatmospheric pressure (iron lung) or has a separate negative-pressure generator (Porta-Lung).
  - Uses: Management of respiratory failure; ventilatory support in patients with decreased pulmonary compliance, significant scoliosis, and severe infections
- ***Wrap ventilators (Poncho, Pneumosuit):***
  - Plastic grid that covers the abdomen and thorax. The wrap is sealed around the patient's wrists, neck and abdomen, or legs. A negative-pressure ventilator creates subatmospheric pressure under the grid and wrap.
  - Provides greater volumes
  - Only used with nocturnal assisted ventilation
- Uses: Beneficial in patients with scoliosis or with sensory deficits
  - Disadvantages: Difficult to don, decreased access to the body by the medical personnel, difficult to turn the patient
- ***Cuirass or chest shell ventilators:***
  - Firm shell that covers the chest and abdomen attached to a negative-pressure ventilator that generates a subatmospheric pressure under the shell
  - It is the only NPBV that can be used during the day for ventilatory support in the seated position.
  - Advantages: The patient can get on and off without assistance.
  - Disadvantages: In insensate patients, it can cause pressure ulcers around the area anterior to the axilla.
  - Not effective in:
    - Patients with complete respiratory paralysis
    - Impairment of pulmonary compliance
    - Patients with apnea
    - Patients with intrinsic lung disease
    - Severe spinal deformity
    - Morbid obesity

MANAGEMENT OF OBSTRUCTIVE SLEEP APNEA

General

- Retropharyngeal muscles relax and block the airway.
- ***Risk factors include*** obesity; neck circumference >17 in.; HTN; narrowed airways; chronic nasal congestion; diabetes mellitus (DM); being male, Black, Hispanic, or Pacific Islander; menopause; family history; smoking; and use of alcohol/sedatives.

Treatment Depends on the Findings on the Sleep Study

- Mild OSA:
  - Lifestyle changes
  - Weight loss
  - Smoking cessation
  - Avoidance of alcohol and tranquilizers/sleeping pills → relax muscles, causing interference with breathing
  - Sleep on the side or abdomen rather than on the back in certain patients with positional OSA.
  - Other patients can benefit from a mandibular advancement device or CPAP.
  - If suffering from nasal congestion, use saline nasal spray. Decongestants used for short term only.

- Moderate to severe OSA:
  - 🕮 **CPAP is the preferred method to treat most patients with moderate OSA, but again depends on the sleep study results.**
  - Oral appliances: An orthodontic splint that brings the mandible and tongue forward is helpful to maintain the hypopharynx open as a long-term resource.
  - CPAP or Autopap might be more effective, but oral appliances may be easier to use.
  - Surgical management: Uvulopalatopharyngoplasty (effective in 50% of cases, but often of temporary benefits), maxillomandibular advancement, tracheostomy, and implants that keep the soft palate open (pillar procedure)
- Use of independently varying IPAP and EPAP ventilators (BiPAP or Auto BiPAP) is very effective in patients with hypercapnia and those with severe disease requiring high pressures or those unable to tolerate CPAP use. The greater the difference between IPAP and EPAP, the greater the inspiratory muscle assistance.
- Custom-molded nasal interfaces may be provided to allow for an adequate fit.
- Portable volume ventilators may be used in morbidly obese patients or patients who require high peak ventilator pressures.
- In certain patients unable to tolerate CPAP treatment, Inspire therapy might be indicated. Inspire is an implanted device that stimulates the branch of the hypoglossal nerve that causes tongue protrusion, so as to move the tongue and keep the airway open during respiration.

INVASIVE VENTILATORY SUPPORT

- Invasive ventilation utilizing tracheal intubation is used when noninvasive methods fail and is contraindicated in a patient with respiratory insufficiency. Tracheal intubation or tracheostomy should be considered when the ABGs show PaO_2 <55 mmHg or PCO_2 >50 mmHg.
- **COPD and restrictive lung disease patients may need intubation for other reasons:**
  - Noninvasive mechanical ventilator does not deliver O_2 adequately due to poor access to oral or nasal routes (i.e., orthopedic conditions [osteogenesis imperfecta, inadequate bite or mouthpiece entry], presence of NGT, or upper airway obstruction).
  - Severe intrinsic pulmonary disease requiring high concentration of inspired oxygen (FiO_2)
  - Inadequate oropharyngeal muscle strength (inability to maintain airway)
  - Uncontrolled seizures or substance abuse causing airway compromise
  - Assisted peak cough flow <160 L/min
  - When mechanical exsufflator is not available or contraindicated
  - Unreliable access to assisted coughing
  - Depressed cognitive status

TRACHEAL INTUBATION WITH TRACHEOSTOMY TUBE

- The choice of tracheostomy tube depends upon the patient and the duration of use.
- Tracheostomy tubes have various features that are important to know:
  - Metal versus plastic material
  - Cuffed versus cuffless tubes
  - Fenestrated versus unfenestrated tubes

METAL VERSUS PLASTIC TRACHEOSTOMY TUBES

- ***Metal (e.g., Jackson, Holinger):***
  - Cuffless, reusable tubes made of stainless steel or silver
  - Cause less local irritation and tissue reaction as compared with plastic
  - May be left in place longer
  - Help to keep the tracheostomy stoma open until the tracheostomy is not needed in patients who breathe spontaneously

- ***Plastic (e.g., Bivona, Shiley, Portex):***
  - Disposable, made of PVC, nylon, silicone, and Teflon
  - Available in single or double cannulated, with/without cuff

CUFFED VERSUS UNCUFFED TRACHEOSTOMY TUBES

- *Cuffed tracheostomy tubes:*
  - Provide a good air seal, protect lower airways from aspiration, and prevent air leaking through the upper airway
  - Patients cannot speak with a cuffed tracheostomy tube while the cuff is inflated.
  - Two types:
    - High pressure and low volume (no longer in use)
    - Low pressure/high volume (conform more to the shape of the trachea and inflate more uniformly)
- *Uncuffed tracheostomy tubes:*
  - Some patients may be able to talk while on mechanical ventilation.
  - Should not be used in patients at risk for aspiration because it provides a loose fit
  - Used after tracheostomy, when a looser fit of the tube on the stoma is needed, or to prevent subcutaneous emphysema
  - Used in patients with increased secretions
  - Should not be used in patients known to aspirate

FENESTRATED VERSUS NONFENESTRATED TRACHEOSTOMY TUBES

- *Fenestrated tubes:*
  - 🕮 Good for patients who are able to speak and require only intermittent ventilatory assistance
  - A continuous inner cannula can be used with an outer fenestrated cannula. The fenestrations should lie within the lumen of the trachea and should not touch the tracheal wall (may develop granulation tissue around the holes and become clogged with secretions).
  - The inner cannula can be attached to a positive-pressure ventilator.
  - When the inner cannula is out and the tube is plugged, the patient can breathe through the fenestrations and is able to phonate. This is possible because the air is directed through the upper respiratory tract.
- *Nonfenestrated tubes:*
  - Used in patients who require continuous mechanical ventilation or are unable to protect the airway during swallowing
  - 🕮 If the patient wants to talk, a one-way talking valve may be used on the tracheostomy tube. These devices open on inhalation and close during exhalation to produce phonation.
- Often complicated by increased secretions and should be used cautiously in patients with marginal respiratory reserve

🕮 TALKING TRACHEOSTOMY TUBES VERSUS SPEAKING VALVES

- *Speaking tracheostomy tubes* (e.g., Portex "Talk" tube, Bivona Fome-Cuf with side-port airway connector, Communi-Trach)
  - Used in alert and motivated patients who require an inflated cuff for ventilation and who have intact vocal cords and the ability to mouth words
  - The distinguishing characteristic of talking tracheostomies is that the cuff remains inflated at all times so the patient is able to speak while maintaining a closed system for ventilation.
  - Talking trachs contain a gas line with an external thumb port. When the thumb port is occluded, gas passes through the larynx via small holes above the inflated cuff, allowing the patient to speak.

  - The quality of speech is altered (e.g., lower pitch, coarser, low whisper) and some patients may not be able to produce adequate voice even with practice. Patients need to speak short sentences (because constant flow through the vocal cords can cause the voice to fade away).
  - The patient requires some manual dexterity and minimal strength to occlude the external port.
- *One-way speaking valves* (e.g., Passy-Muir speaking valve, Olympic Trach-Talk; **Table 9–1**):
  - A Passy-Muir valve (PMV) is the only valve that has a biased, closed position; opens only on inspiration.
  - All the other valves are open at all times until they are actively closed during expiration (when enough force is placed).
  - The air is directed into the trachea and up through the vocal cords, creating speech as air passes through the oral and nasal chamber.
  - Requires less work—opening and closing the valve is not needed
  - 🕮 Indications:
    - Patient alert, awake, and attempting to communicate
    - Medically stable, able to exhale efficiently and completely around the tracheostomy tube and the upper airway
    - Able to tolerate complete cuff deflation
    - Able to tolerate speaking valve trial (see next section)
  - 🕮 Contraindications:
    - Unconscious/comatose patients
    - Vocal cord paralysis-adducted position
    - Inflated tracheostomy tube cuff of any kind
    - Foam-filled cuffed tracheostomy tube (may cause airway obstruction)
    - Severe airway obstruction, laryngeal stenosis, or tracheal stenosis
    - Unmanageable, thick secretions

TABLE 9–1 Characteristics of One-Way Speaking Valves

VALVE[a] (MANUFACTURER)	TYPE	VALVE CHARACTERISTICS
Passy-Muir speaking valve (Passy-Muir, Inc.)	One-way valves, use #005 for tracheostomy use, and #007 for ventilator use (only valve for ventilator)	One-way silastic membrane with biased closed position–opens on inspiration. Creates positive closure feature.
Montgomery (Boston Medical Products, Inc.)	One-way valve	Silicone membrane is hinged; maintains open position but opens more fully upon inspiration, closing upon expiration. Special cough release feature.
Tracoe (Boston Medical Products, Inc.)	Two types of fenestrated inner cannulas, which contain hinged valves	Tracheostomy tube is modified by the placement of an inner cannula that contains a one-way valve.
Kistner one-way valve (Pilling Weck)	One-way valve	Hinged valve maintains open position, opening more fully upon inspiration, closing during expiration.
Olympic "Talk Trach" (Olympic Medical)	T-shaped device that fits on tracheostomy tube and can be attached to T-piece	Spring-loaded valve mechanism maintains open position. Closes upon expiration to direct air into the upper airway.
Hood Laboratories	One-way valve	Valve contains a ball that moves, opening upon inspiration and closing upon exhalation.

[a]All valves must be used with deflated tracheostomy tube cuffs.
Source: From Dikeman KJ, Kanandjian MS. *Communication and Swallowing Management of Tracheostomized and Ventilator Dependent Adults*. Singular Publishing Group, Inc; 1995, with permission.

- Severe risk for aspiration
- Severely reduced lung elasticity. Do not use the speaking valves with COPD patients because they cannot force air out due to lack of lung compliance.
- Do not use during sleep. Also do not use with endotracheal (ET) tubes or other artificial airways.

🕮 SPEAKING VALVE TRIAL

1. 🕮 Cuff must be completely deflated when the PMV is on. Failure to deflate can cause immediate respiratory distress. Uncuffed tracheostomy tube is recommended.
2. Attach pulse oximeter.
3. Assess vitals before, during, and after PMV placement: Oxygen saturation, HR, RR, and so on.
4. Monitor breathing sounds; they should remain the same after PMV placement. Decreased breath sounds or prolonged expiratory phase indicates possible airway obstruction.
5. Patient's responsiveness, color, work of breathing, secretion status
6. Slowly deflate tracheostomy tube cuff and allow patient to adjust to cuff deflation.
7. Proceed with PMV attachment (PMV Instruction Booklet).

TRACHEOSTOMY SUCTIONING

- 🕮 Can have the following complications: Bleeding, infection, atelectasis, hypoxemia, cardiovascular instability (including dysrhythmias and in extreme cases cardiac arrest and death), elevated intracranial pressure, and cause lesions to the tracheal mucosa (Pedersen et al., 2009)
- Recommendations:
  - Suction only when necessary.
  - Suction catheter should occlude less than half the lumen of the ET tube.
  - Insert catheter no further than the carina.
  - Suction no longer than 15 seconds.
  - Continuous suction rather than intermittent suctioning during the individual suction procedure
  - Hyperoxygenate before and after suctioning procedure (reduces the risk of arrhythmias)
  - Avoid saline lavage.
  - Use either closed or open suction systems (closed systems may reduce risk of infection and hypoxemia).
  - Use universal precautions.
  - Use lowest possible suction pressure, usually 80 to 120 mmHg.

Guidelines for Decannulation

- Patients are ready for decannulation when they no longer need mechanical ventilation and can adequately clear airway secretions and have an adequate mental status.
- Patient should be evaluated for aspiration risk.
- Patient should be able to cough secretions out of the tracheal tube.
- Gradual cuff deflation allows weaning from cuffed to uncuffed trach. The cuffless trach is downsized to a smaller size and the patient evaluated for ability to cough secretions.
- When the patient does not need excessive suctioning and the outer diameter of the tracheostomy tube is 8 mm, you may discontinue the trach or place a tracheal button temporarily assuming stability of the underlying medical condition that necessitated the tracheostomy initially.

Tracheal Buttons

- Extend only to the inner surface of the anterior tracheal wall without causing tracheal lumen obstruction

- They are used when there is doubt about the success of the tracheostomy weaning. When plugged, the patient may breathe through the upper airway without resistance from the tracheostomy tube.

🕮 Diaphragmatic Pacer

- Another means of invasive ventilatory support is electrophrenic respiration with the use of a diaphragmatic pacer in patients with intact phrenic nerves and diaphragm. This is discussed in detail in Chapter 7, Spinal Cord Injuries.

CARDIAC REHABILITATION

DEFINITION

- Cardiac rehabilitation (CR) is a medically supervised specialty with a multidisciplinary approach to deliver a comprehensive secondary prevention program. Cardiac rehabilitation optimizes cardiovascular disease (CVD) risk reduction, promotes adherence to healthy lifestyle behaviors, and reduces disabilities. Specific components of CR include medical evaluations, psychosocial assessment, physician-prescribed exercise, cardiac risk factor modification, education, behavioral counseling, and outcomes assessment (American Association of Cardiovascular and Pulmonary Rehabilitation [AACVPR], 2021).

GOALS

- The overall goal is to improve and maintain a good level of cardiovascular fitness, thereby returning the individual to as normal and productive of a life as possible.
  - Eligible patients with acute coronary syndrome (ACS), immediately post-coronary artery bypass grafting (CABG), post-percutaneous coronary intervention (PCI), chronic stable angina, stable chronic heart failure (HF), or who are postcardiac transplantation or cardiac valve surgery should be referred to a comprehensive outpatient cardiovascular rehabilitation program.
- For those able to return to work:
  - Return to productive employment as soon as possible.
  - Improve and maintain good cardiovascular fitness.
- For those not able to return to work:
  - Maintain as active of a life as possible.
  - Establish new areas of interest to improve quality of life.
- Patient education and reduction of coronary risk factors:
  - The educational program must address reversible causes of ischemia.
    - Barring contraindication, all MI survivors should receive a cholesterol-lowering agent, aspirin, and a beta-blocker. Aspirin reduces the incidence of subsequent MI, stroke, and death from cardiovascular causes by about 25%.
    - Appropriate treatment of HTN with afterload reduction, beta-blockade, and use of anti-platelet agents improve overall survival.
- **Core components of cardiac rehabilitation and secondary prevention programs:**
  - **Patient assessment**: Perform medical history and exam, provide appropriate cardiac testing including resting EKG, and ensure medication compliance (acetylsalicylic acid [ASA], clopidogrel, beta-blockers, lipid-lowering agents, angiotensin-converting enzyme [ACE] inhibitors, or angiotensin II receptor blockers [ARBs] as recommended by the American Heart Association/American College of Cardiology [AHA/ACC]).
  - **Nutritional counseling**: Decreased dietary saturated fat and cholesterol content, foods high in sodium and added sugars, individualized plan for addressing comorbidities including obesity, HTN, DM, and CHF

- **Weight management**: Calculating body mass index (BMI; >25 kg/m²) to establish short- and long-term goals; develop combined diet, physical activity, behavioral program designed to reduce total caloric intake; maintain appropriate intake of nutrients and fiber; and increase energy expenditure
- **Blood pressure (BP) management**: High BP should be treated earlier with lifestyle changes and, in select patients, with medications at 130/80 rather than 140/90 (2017 AHA/ACC guidelines; Whelton et al., 2022).
- **Lipid management**: Dietary modifications, lipid-lowering agents in select patients, target treatments toward decreasing low-density lipoproteins (LDL) and triglycerides
- **Diabetes management**: Education of hypo- and hyperglycemia signs and symptoms, blood sugar checks, adequate hydration; tight glucose control can improve outcomes for heart disease.
- **Tobacco cessation**: Assess readiness to change; "5 As" (ask, advise, assess, assist, and arrange); utilizing tobacco cessation programs and pharmacologic interventions in selected patients
- **Psychosocial management**: Individual or small group education and counseling, supportive rehabilitation environment, and community resources
- **Physical activity counseling**: Encourage 30 to 60 minutes per day of moderate-intensity physical activity >5 days per week, advise low-impact aerobic activity, gradual increase in volume of physical activity over time
- **Exercise training**: Individualized exercise prescription for aerobic and resistance training including frequency, intensity, duration, modalities, and progression, including warm-up, cool-down, and flexibility exercises

RISK FACTORS FOR CAD	
NONMODIFIABLE FACTORS	**MODIFIABLE FACTORS**
Age (men ≥45 years old, women ≥55 years old) Male gender Family history of CAD (in male first-degree relative <55 years old, female first-degree relative <65 years old) Past history of CAD, PVD, CVA	Hypertension Cigarette smoking Alcohol or substance abuse Dyslipidemia Diabetes mellitus Obesity/body composition Sedentary lifestyle/physical inactivity Stress or psychological concerns Environmental concerns (e.g., air pollutants)

CAD, coronary artery disease; CVA, cerebrovascular accident; PVD, peripheral vascular disease.
Sources: From American Association of Cardiovascular and Pulmonary Rehabilitation. *Guidelines for Cardiac Rehabilitation and Secondary Prevention Programs*. 6th ed. Human Kinetics; 2021; National Heart, Lung, and Blood Institute. Ischemic heart disease. https://www.nhlbi.nih.gov/health-topics/ischemic-heart-disease.

EPIDEMIOLOGY

- CVD is the leading cause of morbidity and mortality in the United States, accounting for almost 50% of all deaths.
- Coronary heart disease (CHD), with its clinical manifestations as stable angina pectoris, unstable angina, acute myocardial infarction (AMI), CHF, cardiomyopathy, and sudden death, affects about 13.5 million Americans.
- Based on data from the National Health and Nutrition Examination Survey (NHANES) 2017–2020, about 20.5 million Americans ≥20 years of age have CHD (Tsao et al., 2023).
- Estimated annual incidence of MI in America is 805,000, of which about 605,000 are new attacks and 200,000 are recurrent attacks (Tsao et al., 2023).
- 50% of MIs occur in people under age 65.
- Annually, one million survivors of MI and more than seven million patients with stable angina pectoris are candidates for cardiac rehabilitation, as are patients following CABG (309,000 patients in 1993). A similar number will require angioplasty.

- It is projected that in 2018, approximately 720,000 Americans will have a new coronary event (defined as first hospitalized MI or CHD death) and approximately 335,000 will have a recurrent event.
- CHD (43.8%) is the leading cause of deaths attributable to CVD in the United States, followed by stroke (16.8%), high BP (9.4%), HF (9.0%), disease of the arteries (3.1%), and other CVDs (17.9%; Benjamin et al., 2018).
- About 35% of the people who have a coronary event in a given year will die as a result of it (Tsao et al., 2023).
- Although several million patients with CHD are candidates for cardiac rehabilitation services, only 11% to 20% have participated in cardiac rehab programs. Women are referred less to cardiac rehab programs (see recent analysis later).
- An analysis of Medicare claims data revealed only 13.9% of Medicare beneficiaries enroll in cardiac rehabilitation after an AMI and only 31% enroll after CABG. Older people, females, non-Whites, and individuals with comorbidities were less likely to enroll in cardiac rehabilitation programs (Suaya et al., 2007).
- In the National Cardiovascular Data Registry (NCDR) Acute Coronary Treatment and Intervention Outcomes Network (ACTION) Registry-Get With The Guidelines (GWTG), cardiac rehabilitation referral after patients were admitted with a primary diagnosis of ST elevation myocardial infarction (STEMI) or non-ST elevation myocardial infarction (NSTEMI) increased from 72.9% to 80.7% between 2007 and 2012 (Beatty et al., 2014).
- The mortality rate for CAD has fallen 47% since 1963, with 30% of that decrease occurring from 1979 to 1989.
- The Framingham study credits three factors as playing possible roles in this marked decrease in those with CAD (Wilson et al., 1987).
  1. Modification of CAD risk factors:
     - Lower cholesterol
     - Better HTN management
     - Reduced cigarette smoking
  2. Improved treatment methods
  3. Improved prevention

EPIDEMIOLOGIC OUTCOMES

- Survival is improved by cardiac rehabilitation, with patients involved in cardiac rehabilitation after an MI having a 3-year survival rate of 95% compared with 64% in nonparticipants. There is also a significant reduction (28%) in risk of recurrent MI.
- The value of cardiac rehabilitation is supported by a large body of evidence and its recommendation by the AHA/ACC (Bracewell et al., 2022).
- Effects of cardiac rehabilitation on CAD:
  - Significantly lower rates of MI and all-cause hospitalization at 6 to 12 months, and lower rates of MI and cardiovascular mortality at medium-term follow-up (>12–36 months) and long-term follow-up (>36 months).
  - A significantly higher rate of event-free survival (major adverse cardiac events = stroke, target-vessel revascularization, PCI, or CABG) at 12 months.
    - Significant improvement in health-related quality of life at 1 year
    - Significant improvement in clinical symptoms during exercise
    - 20% increase in maximal exercise tolerance
    - Increase in the ischemic threshold (time to signs of ischemia during stress testing including ST depressions and/or angina)
- Effects of cardiac rehabilitation in CHF:
  - Reduced combined rate of cardiovascular mortality or HF hospitalization, increase in cardiopulmonary exercise time, increased peak oxygen consumption, significant improvement in health-related quality of life, significant improvement in 6-minute walk test distance

- Effects of cardiac rehabilitation/supervised exercise therapy in peripheral artery disease (PAD):
  - Increased pain-free walking distance, increased maximal walking distance, improved quality of life
- A community-based analysis in Olmsted County, Minnesota, between 1987 and 2010 revealed an approximately 52.5% participation rate in cardiac rehabilitation after the patients' first MI. After adjustment, cardiac rehabilitation participants had lower all-cause readmission, cardiovascular readmission, noncardiovascular readmission, and mortality risk (Dunley et al., 2014).
- A Cochrane systematic review of 63 studies concluded that exercise-based cardiac rehabilitation programs for CHD patients reduced cardiovascular mortality and hospital admissions (Anderson et al., 2016; Dunley et al., 2014; Hammill et al., 2010).
- One study revealed a strong dose–response relationship between the number of cardiac rehabilitation sessions and long-term outcomes among Medicare beneficiaries, in which attending all 36 sessions reimbursed by Medicare was associated with lower risks of mortality and MI at 4 years compared with attending fewer sessions (Hammill et al., 2010).
- A 45% to 47% decrease in all-cause mortality is seen in patients who participated in cardiac rehabilitation after PCI compared with those who did not participate in cardiac rehabilitation (Goel et al., 2011).
- There is a strong protective association between ideal cardiovascular health metrics and many clinical and preclinical conditions, including premature all-cause mortality, CVD mortality, ischemic heart disease mortality, HF, carotid arterial wall stiffness, coronary artery calcium progression, impaired physical function, cognitive decline, stroke, depression, end-stage renal disease (ESRD), COPD, deep vein thrombosis/pulmonary embolism (DVT/PE), better leukocyte telomere length, and reduced mean healthcare expenditures (Benjamin et al., 2018).

PATHOPHYSIOLOGY

- The response to injury hypothesis considers atherosclerosis to be a chronic, inflammatory response of the arterial wall.
- The initial insult is injury to the endothelium that results in endothelial dysfunction.
- Lipoproteins accumulate in the vessel wall and are oxidized.
- Monocytes attach to the endothelium, migrate into the intima, and transform into foam cells.
- Platelets adhere and are activated to release factors.
- Smooth muscle cells proliferate and contribute to an increase in the extracellular matrix.
- Lipids accumulate in smooth muscle cells and foam cells.
- Fibromuscular plaque results—fibromuscular layer with cholesterol core

PHASES OF CARDIAC REHABILITATION

Phase I	During acute inpatient hospitalization
Phase II	Supervised, outpatient cardiac rehab lasting 3–6 months
Phases III/IV	Maintenance phase in which physical fitness and risk factor reduction are accomplished in a minimally supervised or unsupervised setting

Phase I (Inpatient Period)

- This stage of rehabilitation can last from 1 to 14 days for cardiovascular patients undergoing invasive procedures or suffering from acute events.

Phase II (Immediate Outpatient Period)

- 🕮 This period is the convalescent stage following a hospital discharge. The length is partly determined by risk stratification and monitoring need. By definition, this period is the most closely monitored phase of rehabilitation.

Phase III/IV (Intermediate and Maintenance Periods)

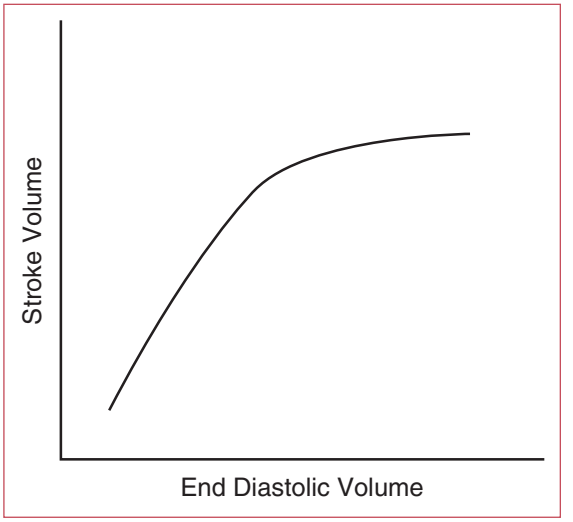
- **Note:** The third stage of cardiac rehab can be broken down into Phases III and IV.
- Phase III of recovery is an extended outpatient period that may be divided into two components: *intermediate* and *maintenance*. The *intermediate stage* follows immediate outpatient cardiac rehabilitation when the patient is not intensely monitored and/or supervised but is still involved in regular endurance exercise training and lifestyle change.
- The transition to Phase IV varies according to the individual outcomes and medical needs.

🕮 EXERCISE PHYSIOLOGY

- **Total oxygen consumption (VO_2)** represents the oxygen consumption of the whole body. It corresponds to the work of the peripheral skeletal muscles rather than myocardial muscles.
- **Aerobic capacity (VO_2max)** is a term used to measure the maximum oxygen consumption that an individual can achieve during exercise. As the individual increases the workload (exercise), the VO_2 increases in a linear fashion until it levels off and reaches a plateau despite further increases in the workload. This is the aerobic capacity of the individual. It is usually expressed in milliliters of O_2 consumed per kilogram of body weight per minute (mL O_2/kg/min).
- VO_2max provides a reliable and reproducible measure of dynamic work capacity as well as cardiovascular fitness.
  - It provides information regarding prognosis in patients with heart disease and can assist in evaluating work resumption after recovery.
  - Treadmill or leg cycle ergometer testing is primarily used to estimate VO_2max (Stein & Brandstater, 2010).
- **Myocardial oxygen consumption (MVO_2)** is the actual oxygen consumption of the heart (i.e., myocardial workload). It can be measured directly with cardiac catheterization. In a clinical setting, however, the **rate pressure product (RPP)** can be used to estimate the MVO_2 since the HR and systolic blood pressure (SBP) correlate well with it (Gobel et al., 1978).
- **Double product**, also called **RPP**, refers to the work required of the heart, which closely parallels the SBP × HR.
  - Estimated MVO_2 = RPP = SBP × HR
- **Cardiac output (CO)** = HR × Stroke volume (SV)
- **Fick equation**: VO_2max = CO × AVO_2 difference
- **Metabolic equivalent (MET):**
  - Defined as the ratio of working metabolic rate to basal (resting) metabolic rate
    - 1 MET = 3.5 mL O_2 consumed per kilogram of body weight per minute (Pashkow, 1993)
  - 1 MET = Energy consumption while at basal metabolic rate (seated rest)

FRANK–STARLING RELATIONSHIP

- The Frank–Starling relationship describes the ability of the myocardium to increase its force of contraction in response to increased stretch (increased ventricular volume or increased preload) placed on it. As a result, SV will increase during the next contraction when venous return is increased. This process is known as the Frank–Starling mechanism (or Stanley's law of the heart). It enables the heart to eject the additional venous return, thereby increasing SV. Graphically represented, this is known as the Frank–Starling curve.



Frank–Starling Relationship.

- Increased venous filling rate and SV increases venous return and ventricular filling (end-diastolic volume) and therefore preload, which is the initial stretching of the cardiac myocyte prior to contraction.
- Myocyte stretching increases the sarcomere length, which causes an increase in force generation. This mechanism enables the heart to eject the additional venous return, thereby increasing SV.

OUTCOMES OF CARDIAC REHABILITATION SERVICES

- The results of cardiac rehabilitation services are based on reports in the scientific literature. The most substantial benefits are the following:

1. Improvement in Exercise Tolerance

- Cardiac rehabilitation exercise training improves objective measures of exercise tolerance in men and women, including elderly patients with CHD and HF.

2. Improvement in Symptoms

- Cardiac rehabilitation exercise training decreases symptoms of angina pectoris in patients with CHD and decreases symptoms of HF in patients with left ventricular (LV) systolic dysfunction. There is also improvement in clinical measures of myocardial ischemia, as identified by EKG and nuclear cardiology techniques following exercise rehabilitation.

3. Improvement in Blood Lipid Levels

- Multifactorial cardiac rehabilitation in patients with CHD, including exercise training and education, results in improved lipid and lipoprotein levels.
- Exercise training as a sole intervention has not affected consistent improvement in lipid profiles.
- Optimal lipid management requires specifically directed dietary and, when medically indicated, pharmacologic management as a component of a multifactorial cardiac rehabilitation program.

4. Reduction of Cigarette Smoking

- Education, counseling, and behavioral intervention are beneficial for smoking cessation during cardiac rehab.

5. Improvement in Psychosocial Well-Being and Stress Reduction

- Improvement in psychological status and functioning, including measures of emotional stress and reduction of the Type A behavior pattern

TABLE 9–2A Medicare-Compliant Diagnostic Categories for Cardiac Rehabilitation Program Services; furnished on or after January 1, 2010

Medicare covers cardiac rehabilitation items and services for patients who have experienced one or more of the following:

- An acute myocardial infarction (within the preceding 12 months); or
- A coronary artery bypass surgery; or
- A current stable angina pectoris; or
- Heart valve repair or replacement; or
- Percutaneous transluminal coronary angioplasty (PTCA) or coronary stenting; or
- A heart or heart-lung transplant; or
- Stable, chronic heart failure defined as patients with left ventricular ejection fraction of 35% or less and New York Heart Association (NYHA) class II to IV symptoms despite being on optimal heart failure therapy for at least 6 weeks (effective February 18, 2014)

Source: From Medicare: Cardiac Rehabilitation Program Coverage. Accessed September 1, 2019. https://www.medicare.gov/coverage/cardiacrehabilitation-programs.

TABLE 9–2B Absolute and Relative Contraindications to Exercise Training

Absolute
• Recent change in the resting EKG suggesting significant ischemia, recent MI, or other acute cardiac event
• Unstable angina
• Uncontrolled cardiac arrythmias
• Symptomatic severe aortic stenosis or other valvular disease
• Decompensated symptomatic heart failure
• Acute pulmonary embolus or pulmonary infarction
• Acute noncardiac disorder that may affect exercise performance or may be aggravated by exercise (e.g., infection, thyrotoxicosis)
• Acute myocarditis or pericarditis
• Acute thrombophlebitis
• Physical disability that would preclude safe and adequate exercise performance
Relative[a]
• Electrolyte abnormalities
• Tachyarrhythmias or bradyarrhythmias
• High-degree atrioventricular block
• Atrial fibrillation with uncontrolled ventricular rate
• Hypertrophic obstructive cardiomyopathy with peak resting left ventricular outflow gradient of >25 mmHg
• Known aortic dissection
• Severe resting arterial hypertension (systolic BP >200 mmHg and diastolic BP >110 mmHg)
• Mental impairment leading to inability to cooperate with testing

[a]Contraindications can be superseded if benefits outweigh risks of exercise.
BP, blood pressure; MI, myocardial infarction.
Sources: Adapted from American Association of Cardiovascular and Pulmonary Rehabilitation. *Guidelines for Cardiac Rehabilitation and Secondary Prevention Programs*. 5th ed. Human Kinetics; 2013; Gibbons J, et al. 2002, ACC/AHA 2002 guideline update for exercise testing. A report of the American College of Cardiology/American Heart Association Task Force on practice guidelines (Committee to Update the 1997 Exercise Testing Guidelines). American College of Cardiology. https://www.ahajournals.org/doi/10.1161/01.cir.0000034670.06526.15

6. Reduction in Mortality

- Multifactorial cardiac rehabilitation service can reduce cardiovascular mortality in patients following MI.

7. Safety

- The safety of exercise is established by the very low rate of occurrence of MI and cardiovascular complications during exercise training.

Parameter Changes in Patients Who Participate in Cardiac Rehabilitation (Whiteson & Einarsson, 2011)

CORONARY ARTERY DISEASE

- Cardiac rehabilitation in patients with CAD is associated with a reduction in morbidity and mortality partly due to better autonomic regulation of the heart.
- Improvement in LV contractile function is also noted.
- Inpatient cardiac rehab for severe angina improves physical performance and improves rates of return to work.

STABLE ANGINA AND MYOCARDIAL INFARCTION

- After a small MI, exercise training is started in 2 to 4 weeks.
- After a large MI, exercise training is deferred to 4 to 6 weeks, starting with a lower intensity level, frequent clinical surveillance, and EKG monitoring.
- After MI, a change in LV dimensions (remodeling) occurs over weeks to months.
  - Remodeling is linked to worsening LV function, ventricular arrhythmias, aneurysm formation, and higher mortality.

- Cardiac rehab also improves myocardial perfusion and LV electrophysiologic parameters, reducing the risk for malignant ventricular arrhythmias and sudden cardiac death after MI.
- Exercise prescription is similar for stable angina and post-MI.
- Following MI, HR increase should be kept within 20 bpm and SBP within 20 mmHg of baseline with activity during the acute phase/Phase I of cardiopulmonary rehabilitation. Target intensity by the end of the Phase I exercise program should be aimed at 4 METs (Bartels & Prince, 2016).

ANGIOPLASTY AND STENTING

- No increase in the rate of stent restenosis with cardiac rehabilitation participation
- Aerobic training programs after angioplasty improve VO_2max and increase functional capacity.
- After angioplasty, patients who completed 6 months of exercise training improved respiratory efficiency and LV systolic function, reduced damaging remodeling, stopped further CHD, and decreased CHF progression.

CORONARY ARTERY BYPASS GRAFT SURGERY

- Improved myocardial perfusion is associated with an increased ischemic threshold.
- Usual exercise physiologic changes occur, with peripheral adaptations leading to improvement in VO_2max.
- Moderate-intensity resistance weight training is safe and effective but does not increase VO_2 without the concomitant use of aerobic exercise.
- Interval training gives a more rapid increase in exercise capacity compared with steady-state aerobic training.
- After CABG, if the ejection fraction (EF) is reduced, then the cardiac rehabilitation program should be adjusted to follow the same protocol as that for CHF patients.

CARDIOMYOPATHY

- Patients with severe ischemic cardiomyopathy (e.g., EF <20%) tolerate cardiac rehab, with increased VO_2max, improved LV systolic and diastolic function, and enhanced quality of life.

CONGESTIVE HEART FAILURE

- A significant increase in VO_2max and exercise tolerance is noted with cardiac rehab for CHF.
- Cardiac rehabilitation for CHF improves myocardial perfusion.
- Cardiac rehabilitation results in fewer cardiac events, significantly decreased rates of hospital readmission for CHF, reduced cardiac mortality, and enhanced quality of life.
- Interval training improves VO_2max and muscle strength.
- Resistance training along with aerobic training shows greater improvements in LV function, increased VO_2max, and strength compared with aerobic training alone.

HEART TRANSPLANTATION

- Cardiac rehabilitation is safe for patients receiving intravenous (IV) inotropic support who are awaiting transplantation and is recommended both before and after heart transplantation (HT).
- A significant increase in VO_2max and work capacity with cardiac rehabilitation is observed.
- Left ventricular assist device (LVAD): Functional status is more significantly improved after HT compared with LVAD insertion.

CARDIAC ARRHYTHMIA

- In one-third of patients, atrial tachyarrhythmias can arise immediately after cardiac surgery.
- Atrial fibrillation (AF or AFib) is the most common tachyarrhythmia and frequently persists during acute inpatient cardiac rehabilitation.

- Roughly 5% of patients doing cardiac rehabilitation after CABG do not complete the cardiac rehabilitation program because of the development of arrhythmias (25% are ventricular).
- Supine exercise is more likely to induce ventricular arrhythmias than upright exercise.
- Patients with an implanted cardioverter-defibrillator (ICD) tolerate an exercise training intensity equal to that for patients without a device, with a significant increase in VO_2max.

INPATIENT VERSUS OUTPATIENT CARDIAC REHABILITATION PROGRAMS

1. Inpatient program: Strictly supervised inpatient hospitalization lasting 1 to 2 weeks (Phase I)
2. Structured outpatient program: Supervised ambulatory outpatient program lasting 3 to 6 months (Phase II)
3. Maintenance program: Minimally supervised or unsupervised setting (Phase III/IV)

Inpatient Program 7 to 14 Days

- The goal of an inpatient rehabilitation program is to provide a coordinated, multifaceted program designed to assist and direct patients and their families early in the recovery process following an acute cardiovascular event.
- The focus is on the medical care, physical activity, education, and psychological issues.

ACUTE PERIOD—CORONARY CARE UNIT

- Mobilization after cardiac surgery must progress as rapidly as possible to prevent deconditioning, decubitus ulcer, pneumonia, and thromboembolism.
- Activities of very low intensity (1–2 METs):
  - Passive ROM (1.5 METs)
  - Upper extremity ROM (1.7 METs)
  - Lower extremity ROM (2.0 METs)
  - Avoid: Isometrics (increases HR), Valsalva (promotes arrhythmia), raising the legs above the heart (can increase preload)
- Use protective chair posture; can reduce the CO by 10%
- Bedside commode (3.6 METs) versus bedpan (4.7 METs)

SUBACUTE PERIOD

- Physical program can vary among institutions. Transfer from the coronary care unit (CCU) to either a telemetry unit or to the medical ward.
- Activities or exercises of intensity (3–4 METs):
  - Calisthenics of known energy cost
  - ROM exercise: Intensity can be gradually increased by increasing the speed and/or duration; may add mild resistance or low weight (1–2 lb.).
  - Early ambulation: Starting in the room and then corridors of the ward, treadmill walking at 0% grade starting at 1 mph and gradually increasing to 1.5, 2, and 2.5 mph as tolerated
- Energy cost of low-grade ambulation:
  - 1 mph (slow stroll) = 1.5 to 2 METs
  - 2 mph (regular slow walk) = 2 to 3 METs
  - Propelling wheelchair = 2 to 3 METs
- Serial progression of the self-care activities should parallel the intensity of the monitored program, particularly with earlier hospital discharge.
- **Bypass surgery** rehabilitation regimen is differentiated into aggressive versus slow to recover (Figure 9–8).

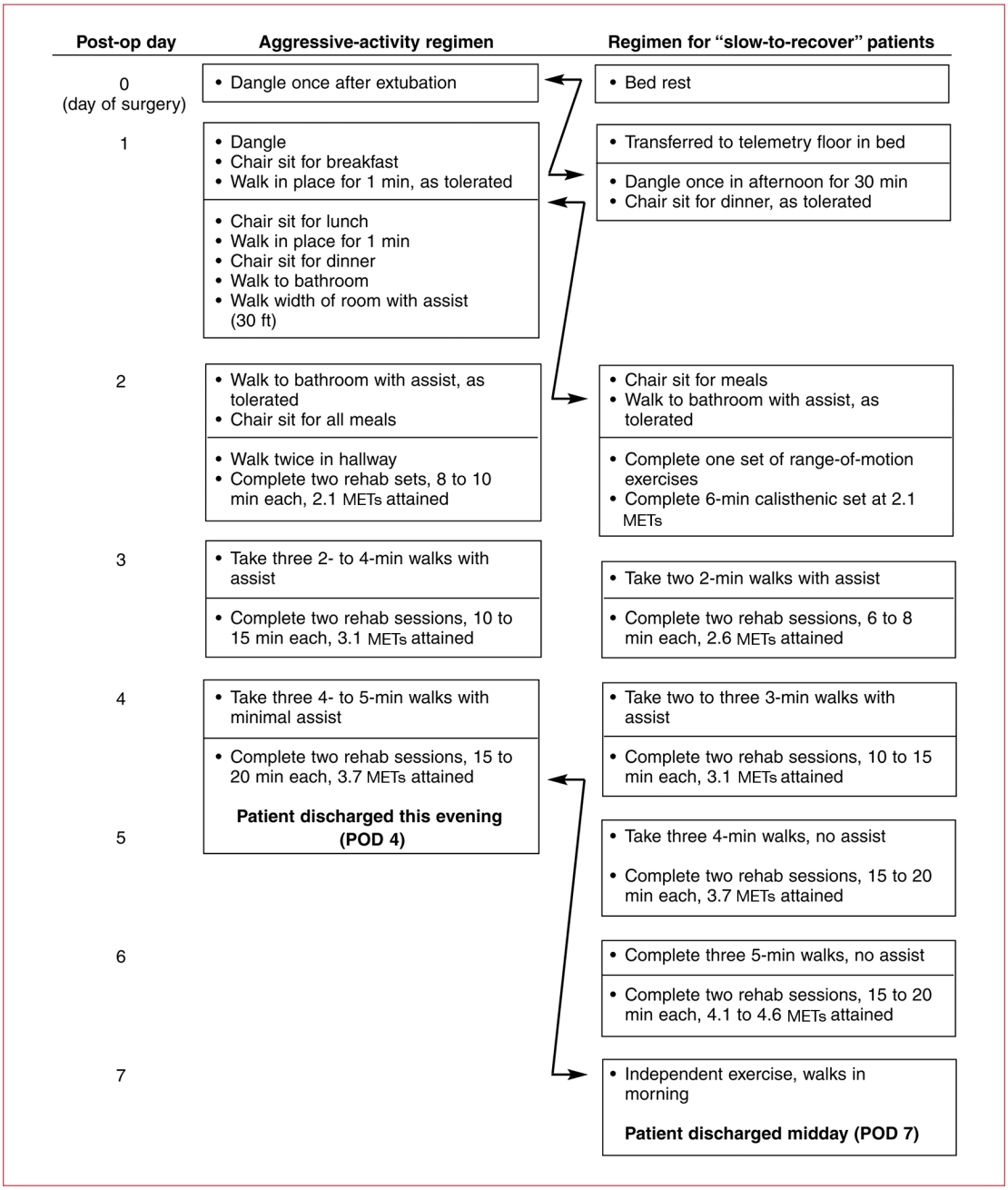


FIGURE 9–8 Activity regimen for patients recovering from cardiac bypass surgery.
MET, metabolic equivalent; POD, post-op day.

EXERCISE TESTING

🕮 Graded Exercise Testing

- Graded exercise stress tests assess the patient's ability to tolerate increased physical stress. The graded exercise testing (GXT) may be used for diagnostic, prognostic, and therapeutic application, with or without addition of radionuclide or echocardiography assessment.
- 🕮 The cardiac rehabilitation health professionals usually use GXTs as functional rather than diagnostic tools.

- 🕮 GXTs also provide useful information when applied to risk stratification models. GXTs also allow the establishment of appropriate limits and guidelines for exercise therapy and the assessment of functional change over time.
- Submaximal GXT is recommended for inpatients and prior to outpatient cardiac rehabilitation programs.
- GXTs may be submaximal or maximal relative to patient effort in addition to common indications for stopping the exercise test (see **Table 9–4**). Endpoint criteria for submaximal testing may include HR limits, perceived exertion, and predetermined MET levels.
- Most of the ADLs in the home environment require less than 4 METs (American Association of Cardiovascular and Pulmonary Rehabilitation, 1995).
- The AHA suggests an HR limit of 30 to 140 bpm for patients not on beta-blocking agents, or a Borg Rating of Perceived Exertion (RPE) Scale of 13 to 15 (see **Table 9–7**), as additional endpoint criteria for low-level testing.
- The low-level test provided sufficient data to permit most ADLs and serves as a baseline for ambulatory exercise therapy.
- The frequency of the test should be relative to the patient's clinical course rather than a fixed schedule.

Exercise Testing With Imaging Modalities

EXERCISE STRESS ECHOCARDIOGRAPHY

- Can be combined with EKG to increase the sensitivity and specificity of stress testing and determine the extent of myocardium at risk for ischemia. Echocardiographic images are obtained while the patient bicycles or immediately after treadmill exercise (images must be obtained within 1–2 minutes after exercise). Myocardial contractibility normally increases with exercise, whereas ischemia causes hypokinesis or dyskinesis of the affected segments. A test is considered positive if wall motion abnormalities develop in previously normal areas with exercise or worsen in an already abnormal segment.
- The overall sensitivity and specificity of exercise echocardiography range from 78% to 97% and 64% to 94%, respectively, with higher sensitivities in patients with multivessel disease. Exercise echocardiography has been shown to be highly accurate in diagnosing CAD in patients in whom there may be an increased incidence of false positive exercise EKG (e.g., women). Stress echocardiography provides an accurate assessment of CAD and yields important diagnostic and prognostic information in the majority of patients.

EXERCISE NUCLEAR STRESS TEST

- Exercise stress tests with nuclear imaging are performed in conjunction with EKG monitoring. Thallium or sestamibi is injected 1 minute prior to exercise and images are obtained. Perfusion defects that are present during exercise, but not seen at rest, suggest ischemia. Perfusion defects that are present during exercise and persist at rest suggest previous MI or scar. In this manner, the extent and distribution of ischemic myocardium can be identified.

PHARMACOLOGIC STRESS TESTING

- Patients unable to undergo exercise stress testing for reasons such as deconditioning, peripheral vascular disease (PVD), orthopedic disabilities, or neurologic disease can benefit from pharmacologic stress testing.
- Two of the most commonly used pharmacologic stress tests:
  - Dobutamine stress echocardiography (DSE)
  - Sestamibi or thallium scintigraphy with dipyridamole or adenosine
- Indications for these tests include establishing a diagnosis of CAD, determining myocardial viability prior to revascularization, assessing prognosis after MI or chronic angina, and evaluating cardiac risks preoperatively. The pharmacologic studies are useful in determining the level of risk stratification, particularly as it relates to the exercise program.

🕮 EXERCISE TESTING PROTOCOLS

- A variety of exercise testing protocols are available, whether the test is conducted using treadmill, cycle, or arm ergometer.
- 🕮 Lower extremity amputee patients can use arm ergometers.
- 🕮 Treadmill testing provides a more common form of physiologic stress (i.e., walking), in which subjects are more likely to attain a slightly higher VO_2max and peak HR.
- 🕮 The cycle ergometer has the advantage of requiring less space and generally is less costly than the treadmill. Minimized movements of the arm and thorax facilitate better quality EKG recording and BP monitoring (American Association of Cardiovascular and Pulmonary Rehabilitation, 1995).
- 🕮 To perform a stress test in an above-the-knee amputee, an upper extremity ergometer is used.
- Balke-Ware protocols that increase metabolic demands by 1 MET per stage are appropriate for high-risk patients with functional capacity of <7 METs.

🕮 Bruce Protocol

- Metabolic demands of >2 METs per stage may be appropriate for low- to intermediate-risk patients with functional capacity >7 METs.
- 🕮 The widely used Bruce Protocol of 2 to 3 METs per stage is useful with stable patients with functional capacities of 10 METs.
- 🕮 Pharmacologic stress testing in debilitated patients for whom exercise testing cannot be performed has been used to evaluate ischemia. The data from pharmacologic testing cannot be used in exercise presumption (**Tables 9–3 to 9–5**; Froelicher, 1987).

Additional Criteria for Stopping Low-Level/Hospital Discharge Exercise Test

1. Exercise heart rate >130 beats per minute
2. Borg RPE (Rating of Perceived Exertion) >15 **(see Table 9–7)**

Suggested Endpoint Criteria for Submaximal Exercise Progress Evaluation

1. Appearance of any criteria that indicate ending an exercise test
2. Exercise heart rate in excess of previous GXT peak heart rate
3. Borg RPE >16

GXT, graded exercise testing; RPE, Rating of Perceived Exertion.
Sources: Modified from Kenney WL, ed. *ACSM's Guidelines for Exercise Testing and Prescription*, 5th ed. Lea & Febiger; 1995, with permission; Fletcher GF, Froelicher VF, Hartley LH, Haskell L, Pollock ML. Exercise standards: a statement for health professionals from the American Heart Association. *Circulation*. 1990;82:2286–2322. doi:10.1161/01.CIR.82.6.2286, with permission.

STRUCTURED OUTPATIENT PROGRAM/MAINTENANCE PROGRAM

Traditionally, outpatient cardiac rehabilitation has been divided into three phases. Statistics on morbidity and mortality for individuals who participate in the different phases differ, based on the phase, diagnosis, demographics, geographical area, comorbidities, and statistical techniques. General trends, based on diagnosis, are included under the Epidemiologic Outcomes section.

– The three phases of outpatient cardiac rehabilitation are:
  - **Phase II (Immediate)** will define the stage of cardiac rehabilitation that occurs immediately after discharge, in which higher levels of surveillance, EKG monitoring, and intensive risk factor modification occur.
  - **Phase III (Intermediate)** is the period of rehabilitation when EKG monitoring occurs only if signs and symptoms warrant, although endurance training and risk factor modification continue.
  - **Phase IV (Maintenance)** is the stage in the program that is structured for patients who have plateaued in exercise endurance and achieved stable risk factor management.

TABLE 9–3 Approximate MET Levels for Simple Exercise Testing Protocols

FUNCTIONAL CLASS	CLINICAL STATUS	O_2 COST ml/kg/min	METS	BICYCLE ERGOMETER	TREADMILL PROTOCOLS									METS
					BRUCE 3 MIN STAGES MPH / %GR	KATTUS MPH / %GR	BALKE-WARE % GRADE AT 3.3 MPH, 1-MIN STAGES	ELLESTAD 3/2/3 MIN STAGES MPH / %GR	USAFSAM MPH / %GR	"SLOW" USAFSAM MPH / %GR	McHENRY MPH / %GR	STANFORD % GRADE AT 3 MPH	STANFORD % GRADE AT 2 MPH	
NORMAL AND I	HEALTHY, DEPENDENT ON AGE, ACTIVITY	56.0	16	1 WATT = 6 KPDS	5.5 / 20		26 25 24 23 22 21 20 19 18 17 16 15 14 13 12 11 10 9 8 7 6 5 4 3 2 1	6 / 15	3.3 / 25					16
		52.5	15		5.0 / 18									15
		49.0	14	FOR 70 KG BODY WEIGHT KPDS 1500		4 / 22		5 / 15			3.3 / 21			14
		45.5	13		4.2 / 16				3.3 / 20					13
		42.0	12	1350		4 / 18					3.3 / 18	22.5		12
	SEDENTARY HEALTHY	38.5	11	1200		4 / 14		5 / 10	3.3 / 15		3.3 / 15	20.0		11
		35.0	10	1050	3.4 / 14					2 / 25		17.5		10
		31.5	9			4 / 10				2 / 20		15.0		9
		28.0	8	900				4 / 10	3.3 / 10		3.3 / 12	12.5		8
	LIMITED	24.5	7	750	2.5 / 12	3 / 10		3 / 10		2 / 15		10.0	17.5	7
II	SYMPTOMATIC	21.0	6	600							3.3 / 9	7.5	14	6
		17.5	5	450	1.7 / 10	2 / 10		1.7 / 10	3.3 / 5	2 / 10	3.3 / 6	5.0	10.5	5
III		14.0	4	300	1.7 / 5				3.3 / 0	2 / 5		2.5	7	4
		10.5	3								2.0 / 3	0.0	3.5	3
		7.0	2	150	1.7 / 0				2.0 / 0	2 / 0				2
IV		3.5	1											1

MET, metabolic equivalent.
Source: From Froelicher VF. *Exercise and the Heart: Clinical Concepts*. 2nd ed. Year Book Medical Publishers; 1987, with permission.

TABLE 9–4 Absolute and Relative Contraindications to Exercise Stress Testing

ABSOLUTE CONTRAINDICATIONS
1. Acute MI (within 2 days) 2. High-risk unstable angina 3. Uncontrolled cardiac dysrhythmias causing symptoms or hemodynamic compromise 4. Active endocarditis 5. Severe symptomatic aortic stenosis 6. Decompensated symptomatic heart failure 7. Acute pulmonary embolus or pulmonary infarction 8. Acute noncardiac disorder that may affect exercise performance or be aggravated by exercise (e.g., infection, renal failure, thyrotoxicosis) 9. Acute myocarditis or pericarditis 10. Physical disability that would preclude safe and adequate test performance 11. Inability to obtain consent
RELATIVE CONTRAINDICATIONS
1. Untreated left main coronary stenosis or equivalent 2. Severe stenotic valvular heart disease 3. Electrolyte abnormalities 4. Tachyarrhythmias or bradyarrhythmias 5. Atrial fibrillation with rapid ventricular rate, such as >150 bpm 6. Hypertrophic cardiomyopathy 7. Mental impairment leading to inability to cooperate with testing 8. High-degree atrioventricular block – Severe resting arterial hypertension (SBP >200 mmHg or DBP >110 mmHg)

bpm, beats per minute; DBP, diastolic blood pressure; SBP, systolic blood pressure; MI, myocardial infarction.
Source: From American Association of Cardiovascular and Pulmonary Rehabilitation. *Guidelines for Cardiac Rehabilitation and Secondary Prevention Programs*. 6th ed. Human Kinetics; 2021.

TABLE 9–5 Indications for Stopping an Exercise Stress Test

ACC/AHA GUIDELINES: INDICATIONS FOR TERMINATING EXERCISE TESTING
Absolute • Drop in SBP >10 mmHg from baseline blood pressure despite an increase in workload, when accompanied by other evidence of ischemia • Moderate to severe angina • Central nervous system symptoms (e.g., ataxia, dizziness, or near-syncope) • Signs of poor perfusion (cyanosis or pallor) • Technical difficulties in monitoring EKG or SBP • Subject's desire to stop • Sustained ventricular tachycardia • Development of bundle branch block that cannot be distinguished from ventricular tachycardia • ST elevation (>–1.0 mm) in leads without diagnostic Q waves (other than V1 or aVR)
Relative • Drop in SBP >10 mmHg from baseline blood pressure despite an increase in workload, in the absence of other evidence of ischemia • ST or QRS changes, such as excessive ST depression (>2 mm of horizontal or downsloping ST segment depression) or marked axis shift • Dysrhythmias other than sustained ventricular tachycardia, including frequent multifocal ectopic beats including ventricular pairs, supraventricular tachycardia, heart block, or bradyarrhythmias • Fatigue, shortness of breath, wheezing, leg cramps, or claudication • Increasing chest pain

ACC, American College of Cardiology; AHA American Heart Association; SBP, systolic blood pressure.
Source: Adapted from Gibbons RJ, Balady GJ, Bricker JT, et al. ACC/AHA 2002 guideline update for exercise testing: summary article. A report of the ACC/AHA task force on practice guidelines (Committee to Update the 1997 Exercise Testing Guidelines). *Circulation*. 2002;106:1883–1892. doi:10.1161/01.CIR.0000034670.06526.15, with permission.

– Medicare (Part B) covers outpatient cardiac rehabilitation programs if any one or more of the following conditions or procedures were present within the prior 12 months:
  - A heart attack
  - Coronary artery bypass surgery
  - Current stable angina
  - A heart valve repair or replacement
  - A coronary angioplasty or coronary stenting
  - A heart or heart-lung transplant
  - Stable chronic HF

  Source: https://www.medicare.gov/coverage/cardiac-rehabilitation.

Common Activities in Early Cardiac Rehabilitation

ACTIVITY	METHOD	MET LEVEL
Toileting	Bedpan, commode, urinal (bed and standing)	1.5–2.5
Bathing	Bed bath, tub bath, shower	1.5–2
Walking	Flat surface 2 mph 2.5 mph 3 mph	 2–2.5 2.5–2.9 3–3.3
Upper body exercise (low to moderate effort, no resistance)	While standing, arms and trunk	2.5–3
Stair climbing	Climbing one flight = 12 steps	3–4

MET, metabolic equivalent.

🕮 Physical Activity Program

ACTIVITY	SPEED	MET LEVEL
Slow walk	2 mph	2–3
Regular speed walk	3 mph	3–4
Brisk walk	3–5 mph	4–5
Very brisk walk	4 mph	5–6
Sexual intercourse[a]	–	3–4
Outdoor work—shovel snow, spade soil	–	7
Jog, walk	5 mph	9
Mop floor	–	2–4
Push power lawn mower	–	4

[a]MET level for sexual intercourse varies depending upon reference source. Tardif (1989) states that patients who reach 5 to 6 METs on stress testing without ischemia or arrhythmias can, in all likelihood, resume their normal sexual activities without any risk.
MET, metabolic equivalent.
Source: From American Association of Cardiovascular and Pulmonary Rehabilitation. *Guidelines for Cardiac Rehabilitation and Secondary Prevention Programs*. 5th ed. Human Kinetics; 2013.

SPORT ACTIVITY	ENERGY COST IN METS
Golf	2–5
Bowling	4–5
Volleyball	3–4
Ping pong	3–6
Tennis	4–7
Roller-skating	5–6

METs, metabolic equivalents.

- The goal is the improvement of the cardiovascular capacity through physical exercise training whether in a minimally supervised or unsupervised setting.
- Types of physical activities:
  - Begin with the last exercise program performed during the supervised cardiac exercise program.
  - Aerobically trained, clinically stable candidates may participate in resistive or circuit training. An overall lifestyle that includes proper diet, weight control, stress management, and smoking cessation should be maintained along with good physical fitness.
  - Active participation, within prescribed limits, in sport activity is encouraged.
- 🕮 Sexual intercourse is a special consideration, as there can be potential for triggering another cardiac event.
  - It is not recommended following an MI for 2 weeks.
  - Intercourse is as physically intense as climbing two flights of stairs.
  - Intercourse with familiar partners in a known environment requires 4 METs.
  - Achieving 6 METs on a stress test without event indicates low risk for a cardiac event during sexual intercourse.
  - Slow progression in reestablishing a romantic relationship to foreplay and then more physically demanding intimacy is encouraged.
- Reassure that sexual activities with CAD are relatively safe (Whiteson & Einarsson, 2011).

NEW YORK HEART ASSOCIATION CARDIAC FUNCTIONAL CLASSIFICATION

New York Heart Association Class I

- Patient's cardiac disease does not limit physical activity. Ordinary physical activity does not cause undue fatigue, palpitation, dyspnea, or anginal pain.
- Specific Activity Scale:
  - Patients can perform to completion any activity requiring ≥7 METs:
    - Can carry 24 lb. up eight steps
    - Can carry objects that weigh 80 lb.
    - Do outdoor work (shovel snow, spade soil)
    - Do recreational activities (skiing, basketball, squash, handball, jog at 5 mph)

New York Heart Association Class II

- Patient's cardiac disease results in slight limitation on physical activity. They are comfortable at rest. Ordinary physical activity results in fatigue, palpitation, dyspnea, or anginal pain.
- Specific Activity Scale:
  - Patient can perform to completion any activity requiring ≥5 METs but cannot perform to completion activities requiring ≥7 METs:
    - Sexual intercourse to completion without interruption

- Garden, rake, weed
- Roller-skate, walk at 4 mph on level ground

New York Heart Association Class III

- Patient's cardiac disease results in marked limitation of physical activity. They are comfortable at rest. Less than ordinary physical activity causes fatigue, palpitation, dyspnea, or anginal pain.
- Specific Activity Scale (Table 9–6):
  - Patient can perform to completion any activity that requires ≥2 METs but <5 METs:
    - Shower without interruption
    - Strip and make bed
    - Clean windows
    - Walk 2.5 mph
    - Bowl, golf
    - Dress without stopping

New York Heart Association Class IV

- Patient's cardiac disease results in the inability to carry on any physical activity without discomfort. Symptoms of cardiac insufficiency or angina may be present even at rest. If any physical activity is undertaken, discomfort is increased.
- Specific Activity Scale:
  - Patient cannot or does not perform to completion activities requiring ≥2 METs. Cannot carry out activities in Class I to III.

TABLE 9–6 Criteria for Determination of the Specific Activity Scale Functional Class

	ANY YES	NO
1. Can you walk down a flight of steps without stopping (4.5–5.2 METs)?	Go to #2	Go to #4
2. Can you carry anything up a flight of eight steps without stopping (5–5.5 METs)? Or can you a. have sexual intercourse without stopping (3–4 METs); b. garden, rake, weed (5.6 METs); c. roller skate, fox-trot (5–6 METs); or d. walk at a 4 mph rate on level ground (5–6 METs)?	Go to #3	Class III
3. Can you carry at least 24 lb. up eight steps (10 METs)? Or can you a. carry objects that are at least 80 lb. (8 METS); b. do outdoor work—shovel snow, spade soil (7 METs); c. do recreational activities such as skiing, basketball, touch football, squash, handball (7–10 METs); or d. jog/walk 5 mph (9 METs)?	Class I	Class II
4. Can you shower without stopping (3.6–4.2 METs)? Or can you a. strip and make a bed (3.9–5 METS); b. mop floors (4.2 METs); c. hang washed clothes (4.4 METs); d. clean windows (3.7 METs); e. walk 2.5 mph (3–3.5 METs); f. bowl (3–4.4 METs); g. play golf (walk and carry clubs) (4.5 METs); or h. push a power lawn mower (4 METS)?	Class III	Go to #5
5. Can you dress without stopping because of symptoms (2–2.3 METs)?	Class III	Class IV

MET, metabolic equivalent.
Source: From Goldman L, Hashimoto B, Cooke F, Loscalzo A. Comparative reproducibility and validity of systems for assessing cardiovascular functional class: advantages of a new specific activity scale. *Circulation*. 1981;64(6):1227–1234. doi:10.1161/01.CIR.64.6.1227. © American Heart Association, with permission.

EXERCISE PRESCRIPTION FOR CARDIAC PATIENTS

Exercise Prescription

- Exercise for the cardiac patient should specify the type of exercise, the intensity, duration, and frequency.

TYPES OF EXERCISE

- Exercise for cardiovascular conditioning should be isotonic, rhythmic, and aerobic; should use large muscle masses and should not involve a large isometric component.
- Sessions of exercise should incorporate aerobic activity, such as walking/jogging, stationary cycling, or water aerobics. Sessions should also incorporate warm-up and cool-down periods. In addition to aerobic activity, resistance exercise (using lightweights) may be added on an individual basis.
- Resistance exercises have been shown to be a safe and effective method for improving strength and cardiovascular endurance in low-risk patients. Surgical and MI patients should wait 3 to 6 weeks before beginning resistance training.
- Patients diagnosed with the following conditions should be excluded from resistance training:
  - CHF
  - Uncontrolled arrhythmias
  - Severe vascular disease
  - Uncontrolled HTN
    - SBP >160 mmHg or diastolic blood pressure (DBP) >100 mmHg
  - Aerobic capacity <5 METs
- Results in increase in aerobic capacity of all muscle fibers exercised: Both Type I and Type II fibers. Type I fibers continue to show approximately five times the aerobic capacity of Type II fibers, as before exercise (Flores & Zohman, 1993).

EXERCISE INTENSITY

- Exercise intensity is usually prescribed as some percentage of the maximum capacity obtained on exercise testing (i.e., O_2 consumption, HR workload, and/or degree of exertion).

O_2 CONSUMPTION

Threshold	40%–50% max VO_2	60% max HR
Optimum	55%–65% max VO_2	70% max HR
Ceiling	80%–90% max VO_2	90% max HR

HR, heart rate.

- For the deconditioned cardiac patient, exercise even at 40% to 50% of max VO_2 will result in improvement.

TARGET HEART RATE

- Exercise intensity is based on a patient's target HR, which is the HR range based on the clearance HR. There are three main methods of measuring target HR:
  A. Clearance HR method
  B. Age-predicted method
  C. Karvonen method
- Target HR is defined as 70% to 85% of the maximum HR:
  - 0.70 × maximum HR = beginning range of target HR
  - 0.85 × maximum HR = end range of target HR

- **Note: Clearance HR** is the clinical maximum HR attained on stress test.
  A. For the *cardiac patient*, the maximum HR is obtained based on maximum HR achieved on the exercise stress test (clearance HR):
     - 70% of maximum HR attained on exercise stress test
  B. For *healthy patients*, the maximum HR is based on the **age-predicted formula**:
     - **Age-predicted method: 220 – Age = Maximum HR**
     - A simple formula to estimate a maximum HR for a given age. Range from 70% to 85% of predicted adjusted maximum HR.
     - Has the potential for over- and underestimating the actual exercise intensity
     - Can place patients with heart disease at risk for exercise-induced cardiovascular complications
  C. The **Karvonen method** uses the subject's potential HR increase and assumes that the resting HR represents zero intensity:
     - Target HR = 0.70 to 0.85 (max HR – resting HR) + resting HR
     - Corrects for the nonzero value of resting HR
     - Useful for those on chronic beta-blockade or with abnormally high resting HR
     - **Example:** 60-year-old patient with an HR maximum of 160 and a resting HR of 60 bpm
     1. Age-predicted method: Target HR = 0.70 (220 – age) to 0.85 (220 – age)
        Target HR = (220 – 60) × 0.70 to (220 – 60) × 0.85
        Therefore, Target HR = 112 to 136 bpm
     2. Karvonen method: Target HR = 0.7 to 0.85 (max HR – resting HR) + resting HR
        (160 – 60) × 0.7 + 60 = 130 bpm for lower limit
        (160 – 60) × 0.85 + 60 = 145 bpm for upper limit
        Therefore, Target HR = 130 to 145 bpm

RATING OF PERCEIVED EXERTION

1. **Borg RPE Scale (Table 9–7)**:
   Linear scale of rating from 6 to 20. This scale is a valid indication of physical exertion and correlates linearly with HR, ventricular O_2 consumption, and lactate levels. The new exerciser can proceed with exercise to level 13 (somewhat hard) provided they have been given clearance to do so from the exercise stress test.
   – The AHA suggests an HR limit of 130 to 140 bpm for patients not on beta-blocking agents or Borg RPE of 13 to 15 as an additional point criteria for low-level testing.

TABLE 9–7 Borg Scale of Rating of Perceived Exertion

SCALE: 15-GRADE RATING OF PERCEIVED EXERTION
6 No exertion at all
7
8
9 Very light
10
11 Light
12
13 Somewhat hard
14
15 Hard (heavy)
16
17 Very hard
18
19 Extremely hard
20 Maximal exertion

Source: From Borg G. *An Introduction to Borg's RPE Scale*. Movement Publications; 1985, with permission.

2. **Talk test:** Informal and subjective method of estimating exercise level.
   The patient is exercised to an adequate intensity to induce a training effect but is still able to hold a conversation without becoming excessively out of breath.
3. **Patient rating scales (Table 9–8)**:
   – Angina
     - Dyspnea
   – Intermittent claudication

TABLE 9–8 Patient Rating Scales: Angina, Dyspnea, and Intermittent Claudication

ANGINA	DYSPNEA	CLAUDICATION
0—No angina 1—Light, barely noticeable 2—Moderate, bothersome 3—Severe, very uncomfortable 4—Most pain ever experienced	0—No dyspnea 1—Mild, noticeable 2—Mild, some difficulty 3—Moderate difficulty, but can continue 4—Severe difficulty, cannot continue	0—No claudication pain 1—Initial, minimal pain 2—Moderate, bothersome pain 3—Intense pain 4—Maximal pain, cannot continue

Sources: Adapted from American Association of Cardiovascular and Pulmonary Rehabilitation. *Guidelines for Cardiac Rehabilitation and Secondary Prevention Programs*. 3rd ed. Human Kinetics; 1999; American Association of Cardiovascular and Pulmonary Rehabilitation. *Guidelines for Cardiac Rehabilitation Programs*. 2nd ed. Human Kinetics; 1995, with permission.

Duration and Frequency of Exercise

HEALTHY INDIVIDUALS (ACSM 11TH ed., Liguori et al., 2022)

- The American College of Sports Medicine (ACSM) recommends that in order to develop and maintain comprehensive physical fitness, healthy adults need to engage in:
  - Moderate cardiorespiratory aerobic exercise, vigorous cardiorespiratory aerobic exercise, or a combination of moderate-vigorous cardiorespiratory aerobic exercise. (See further details later.)
  - Resistance training exercise. (See further details later.)
  - Flexibility exercise. (See further details later.)
  - Adults can benefit from physical activity of any amount, especially for previously sedentary adults. Physical activity combined in small durations of less than 10 minutes can have positive health outcomes.
- **Cardiorespiratory aerobic exercise:**
  - **Frequency:** ≥3 days per week; spreading sessions over 3 to 5 days a week may be the best way to achieve recommended amount of physical activity.
  - **Intensity**: Moderate (40%–59% of heart rate reserve [HRR]) and/or vigorous (60%–84% of HRR) intensity is recommended for most adults; light- to moderate-intensity exercise may be beneficial in deconditioned persons (**Table 9–9**).
  - **Time:** 30 to 60 minutes per day (150 minutes per week) of purposeful moderate exercise, or 20 to 60 minutes per day (75 minutes per week) of vigorous exercise, or an equivalent combination of moderate and vigorous exercise per day is recommended for most adults.
  - **Type**: Aerobic exercise that involves major muscle groups and is continuous or intermittent in nature is recommended.
  - **Volume:** Target volume of ≥500 to 1,000 MET-minutes per week for most adults is recommended; pedometer step counts of at least 3,000 steps per day at a brisk pace with a minimum daily total of 7,000 to 8,000 steps per day is a reasonable daily goal associated with health benefits.
  - **Progression:** A gradual progression of exercise volume by adjusting exercise duration, frequency, and/or intensity is reasonable until the desired exercise goal (maintenance) is attained. For the average adult, it is reasonable to increase session by 5 to 10 minutes every 1 to 2 weeks for the first 4 to 6 weeks of the exercise program. Downward adjustments are to be made if there are issues with exercise tolerance.
- **Resistance exercise training:**
  - Each major muscle group should be trained at least 2 days per week.
  - At least one set of 8 to 12 repetitions for all major muscle groups is recommended to improve strength and power in most adults.
  - If time is not limited, longer rest intervals (>2 minutes) may allow for a greater amount of work and subsequently greater improvements in muscular fitness.
- **Flexibility exercises:**
  - Used to improve joint-specific ROM and/or to improve activity performance
  - Should be performed ≥2 to 3 days per week, with the greatest gains occurring with daily exercises
  - Holding a static stretch for 10 to 30 seconds is recommended for most adults; in older persons, holding a stretch for 30 to 60 seconds may provide greater benefit.

TABLE 9–9 Methods of Estimating Intensity of Cardiorespiratory Exercise

	CARDIORESPIRATORY ENDURANCE EXERCISE										
	RELATIVE INTENSITY				INTENSITY (%$\dot{V}O_{2MAX}$) RELATIVE TO MAXIMAL EXERCISE CAPACITY IN MET			ABSOLUTE INTENSITY	ABSOLUTE INTENSITY (MET) BY AGE		
Intensity	%HRR or %$\dot{V}O_2R$	%HR_{max}	%$\dot{V}O_{2max}$	Perceived exertion (rating on 6–20 RPE Scale)	20 METs %$\dot{V}O_{2max}$	10 METs %$\dot{V}O_{2max}$	5 METs %$\dot{V}O_{2max}$	METs	Young (20–39 years)	Middle age (40–64 years)	Older (≥65 years)
Very light	<30	<57	<37	Very light (RPE <9)	<34	<37	<44	<2.0	<2.4	<2.0	<1.6
Light	30–39	57–63	37–45	Very light to fairly light (RPE 9–11)	34–42	37–45	44–51	2.0–2.9	2.4–4.7	2.0–3.9	1.6–3.1
Moderate	40–59	64–76	46–63	Fairly light to somewhat hard (RPE 12–13)	43–61	46–63	52–67	3.0–5.9	4.8–7.1	4.0–5.9	3.2–4.7
Vigorous	60–89	77–95	64–90	Somewhat hard to very hard (RPE 14–17)	62–90	64–90	68–91	6.0–8.7	7.2–10.1	6.0–8.4	4.8–6.7
Near-maximal to maximal	≥90	≥96	≥91	≥ Very hard (RPE ≥18)	≥91	≥91	≥92	≥8.8	≥10.2	≥8.5	≥6.8

HR_{max}, maximal heart rate; HRR, heart rate reserve; MET, metabolic equivalent; RPE, Rating of Perceived Exertion; $\dot{V}O_{2max,}$ maximal volume of oxygen consumed per unit time; $\dot{V}O_2R$, oxygen uptake reserve.
Source: From Liguori G, Feito Y, Fountaine C, Roy B. *ACSM's Guidelines for Exercise Testing and Prescription*. 11th ed. Wolters Kluwer; 2022, Table 5.2.

  - A total of 90 seconds of intermittent flexibility exercise per joint is recommended.
  - Joint-specific flexibility exercises are most effective when the muscle is warm.
  - A series of flexibility exercises for each of the major muscle-tendon units is recommended.
- Exercise intensity can be used to modify both cardiopulmonary (aerobic) and resistance training exercises to improve cardiorespiratory fitness.
- The overload principle of training acknowledges that exercising above a minimum intensity or threshold is required to adequately challenge the body to result in changes in physiologic parameters.
  - Highly trained individuals may need to exercise at near-maximal training intensities (i.e., 95%–100% $\dot{V}O_2$max) to improve $\dot{V}O_2$max.
  - Moderately trained individuals may need to exercise at near-maximal training intensities (i.e., 70%–80% $\dot{V}O_2$max) to improve $\dot{V}O_2$max.
- While both moderate- and vigorous-intensity exercise can be used to meet physical activity guideline recommendations, vigorous-intensity exercise (>6 METs; 60%–84% HRR) is more effective at increasing $\dot{V}O_2$max than moderate-intensity exercise (3.0–5.9 METs; 40%–59% HRR), thereby improving cardiorespiratory fitness.
- Interval training yields physiologic adaptations similar to endurance training despite a lower total workload.
- Interval training may include components of both aerobic exercise and resistance training.
- Detrained individuals may benefit from an interval walking training protocol.

Format of an Exercise Session

- Warm-up limited to <15 minutes of light- to moderate-intensity activities
  - Activities specific to muscle groups to be used during exercise to improve ROM
  - May reduce the risk of injury afterwards
  - Dynamic warm-up superior to static flexibility exercises
- Conditioning with 10 to 60 minutes of aerobic, resistance, flexibility, and/or sports activities
- Cool-down with 5 to 10 minutes of light- to moderate-intensity activities
  - At the cool-down period, there is a gradual reduction in exercise intensity to allow the gradual redistribution of blood from the extremities to other tissue and to prevent sudden reduction in venous return, thereby reducing the possibility of postexercise hypotension or even syncope (American Association of Cardiovascular and Pulmonary Rehabilitation, 1995).

Predicted Age-Adjusted Maximum Heart Rate (Figure 9–9)

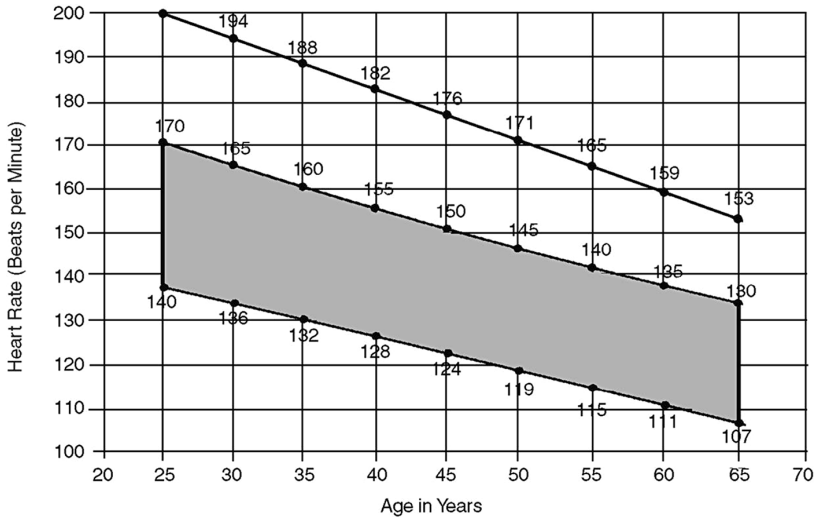


FIGURE 9–9 Exercise intensity based on the predicted age-adjusted rate. Shaded area is 70% to 80% of age-adjusted maximum attainable heart rate. Top solid line is age-adjusted maximum attainable heart rate.

CARDIAC REHABILITATION OF SPECIAL GROUPS

Arrhythmias

- Standard telemetry should be monitored in a cardiac rehab program as noted in phases of cardiac rehabilitation. Warm and cool-down periods decrease the frequency of arrhythmia by promoting coronary perfusion.
- Arrhythmias are frequently noted in patients with CHD after an MI or CABG and are also associated with cardiomyopathy.
- After MI or CABG, the presence of frequent premature ventricular contractions (PVCs) identifies patients at increased risk of developing ventricular tachycardia or sudden cardiac death. Patients with CHD and reduced left ventricular ejection fraction (LVEF) and ventricular tachycardia are more prone to develop ventricular fibrillation.
- Pharmacologic management includes nitrates, beta-blockers, and amiodarone.
- Ablative therapy or an ICD can be considered for life-threatening ventricular tachycardia:
  - ICD decreases mortality by 31% in patients with nonsustained ventricular tachycardia after MI complicated by reduced EF, compared with pharmacologic management alone.
- Nearly 80% of patients with a history of ventricular arrhythmia will have a ventricular arrhythmia during inpatient cardiac rehabilitation. Life-threatening arrhythmias occur in 35% of patients.
- Patients with malignant ventricular arrhythmia, minimally reduced EF, and very good exercise tolerance are more likely to experience ventricular tachycardia or ventricular fibrillation during cardiac rehabilitation than patients with very limited exercise capacity.
  - Patients should decrease exercise intensity and increase exercise duration and frequency. Target HR should be set at a level below which ventricular arrhythmias are noted.
  - The exercise stress test on antiarrhythmic medications is essential to screen for ventricular arrhythmia and to establish cardiac rehabilitation parameters.

Heart Transplantation

- For patients with end-stage heart disease not remediable by more conservative treatment, cardiac transplantation is the accepted form of therapy.
- 🕮 **Orthotopic heart transplantation (OHT)** consists of over 99% of all cardiac transplants:
  - Orthotopic bicaval technique is the preferred method and makes up 75% of OHTs:
    - The donor heart is excised with intact right atrium and a long segment of the superior vena cava.
    - The donor left atrium is sutured to the stump of the four pulmonary veins in the recipient.
    - The superior and inferior venae cavae are sutured to the recipient atrial cuff. The great arteries are anastomosed (Grande et al., 1996).
- 🕮 **Heterotopic transplantation** constitutes less than 1% of transplantations. Here, the recipient heart is left in place to assist the donor heart.
- Epidemiology of cardiac transplants in the United States (Bentley & Ortner, 2020):
  - 3,499 single-organ cardiac transplants performed in the United States in the year 2020
  - Survival rates by year of cardiac transplant in 2020: 90% 1 year, 85% 3 years, and 77% 5 years
  - Majority of transplants (40%) are 50 to 64 years old
  - Male recipients 69% and female recipients 31%
  - 2019 Recipient race/ethnicity for cardiac transplant: White (63%), Black (21%), Hispanic (11%), Asian (3%), Other (2%)
- Indications for cardiac transplant: Cardiomyopathy 60%, CAD 26%, and congenital heart disease 10% (Bentley & Ortner, 2020)

- **Transplant recipient status:**
  - *Status 1A*: These patients are at the top of the waiting list. Includes patients in the ICU, those on life support and/or high-dose IV medications to support their heart function, those who have had a ventricular assist device (VAD), or those who use extracorporeal membrane oxygenation (ECMO) to support their heart function.
  - *Status 1B*: These patients have end-stage HF and are at home on a VAD or continuous IV heart medication (inotrope medication) that makes the heart beat stronger.
  - *Status 2*: These patients do not meet the criteria for Status 1A or 1B. Often these patients are waiting at home for a donor heart and are taking oral HF medication.
  - *Status 7*: These patients are temporarily inactive on the heart transplant waiting list. Includes patients who develop an infection and cannot undergo transplant surgery, those who have left the area and cannot return within 2 hours, or people whose insurance has changed and need a new authorization or have loss of insurance coverage.
- The age range of transplant recipients is from newborn to the eighth decade of life. After transplantation, most patients report a favorable quality of life. Many patients return to work, school, or their usual vocational activities. Typical cardiac transplant patients are middle-aged and have suffered months of preoperative deconditioning, general muscle weakness, depression, and anxiety. A comprehensive rehabilitation approach is necessary.
- Pretransplant management of the patient includes secondary prevention programs and exercise conditioning programs. Any of the several models of LVAD and IV inotropes sustain cardiac function and allow mobilization of these patients.

🕮 Physiologic Response After Heart Transplant

- Posttransplant cardiac rehabilitation programs should focus on conditioning, education, and secondary prevention.
- Physiologic response of the transplanted heart differs from that seen in a post-CABG patient. Transplanted hearts are denervated and lack vagal innervations and central regulation, resulting in the lack of parasympathetic tone due to the lack of vagal inhibition to the sinoatrial (SA) node, resulting in resting tachycardia of 100 to 110 bpm.
- Loss of sympathetic innervation results in circulating catecholamines acting on the chronotropic response, leading to a delayed HR response to exercise.
- Peak HR is typically 20% to 25% lower than controls.
- Patients also have HTN secondary to renal effects of calcineurin inhibitors and prednisone from maintenance medication regimens; diastolic dysfunction may be present in some patients.
- The previously mentioned physiologic conditions result in reduced maximal work output and oxygen by approximately one-third compared with controls (Bartels & Prince, 2016).
- Catecholamine-induced inotropy is reported during peak dynamic exercise in denervated patients (Savin et al., 1980):
  1. High resting HR due to parasympathetic denervation
  2. Lower peak exercise HR
  3. Resting HTN is common, caused in part by the renal effects of anti-rejection medications.
  4. Slower return to resting HR postexercise
  5. At maximum effort, the work capacity, CO, SBP, and the total O_2 consumption (VO_2) are lower.
  6. Pretransplantation rehabilitative strength training may enhance preoperative and postoperative recovery.
  7. Data of survival rates after HT from the registry of the International Society for Heart and Lung Transplantation: 1 year (84.5%), 5 years (72.5%), and 20 years (21%); in some experienced centers, long-term survival is reported to be even higher at 55.6% (Moayedi et al., 2019).
  8. Accelerated atherosclerosis occurs following transplantation.

- Exercise capacity is below normal for most patients, with a mean peak oxygen uptake (VO_2) of approximately 60% of expected (Squires & Bonikowske, 2022).
- Cardiac transplant recipients have abnormal exercise physiology findings related to surgical cardiac denervation, diastolic dysfunction, reduced skeletal muscle oxidative capacity, and impaired vasodilatory ability resulting from pre-cardiac transplant chronic HF (Squires & Bonikowske, 2022).
- Cardiac reinnervation resulting in partial normalization of the HR response to exercise occurs in approximately 40% of cardiac transplant recipients months to years after cardiac transplant (Squires & Bonikowske, 2022).
- Supervised exercise training in cardiac rehabilitation programs is safe and is recommended by professional societies both before (prehabilitation) and after cardiac transplant (Squires & Bonikowske, 2022).
- Exercise training in adults after cardiac transplant improves peak VO_2 and skeletal muscle strength, while also reducing the severity of cardiac allograft vasculopathy (Squires & Bonikowske, 2022).

🕮 Exercise Prescription

- Standard HR guidelines are not used.
- Intensity of exercise is based on the following:
  - Borg RPE Scale 11 to 14 (see **Table 9–7**)
  - Percentage of VO_2max or maximum workload performed on stress test
  - Anaerobic threshold
  - Duration, frequency, and types of exercise follow the same principles as those with other types of cardiac problems.
  - During exercise testing, heart transplant patients with cardiac ischemia do not present with typical symptoms of angina. Instead, EKG changes and other symptoms should be followed.

Outcome

- Generally favorable. Patients typically report increased work output and improved exercise tolerance.

PERIPHERAL ARTERIAL DISEASE

- PAD and CHD often coexist. PAD is present in up to 10 million people in the United States.
- In the United States, 20% of people over age 70 are affected with PAD.
- Intermittent vascular claudication is a symptom of muscle pain, cramping, numbness, and/or fatigue caused by vascular insufficiency from PAD.
  - It is present in up to 40% of patients with PAD.
- 🕮 The ankle–brachial index (ABI) is a ratio comparing the SBP at the ankle to the brachial SBP (ankle SBP/brachial SBP) and is used to classify the severity of PAD.

PAD CLASSIFICATION	
PAD Severity	ABI
Normal	>1.0
Mild	0.8–1.0
Moderate	0.6–0.8
Severe	0.4–0.6
Very severe	<0.4

ABI, ankle-brachial index; PAD, peripheral arterial disease.

Exercise Prescription for Peripheral Arterial Disease and Intermittent Vascular Claudication

- Training intensity that produces moderate claudication pain within the first 5 minutes of treadmill walking. The walking is interspersed with rest periods long enough for symptoms

to subside. Exercise is continued in an exercise–rest interval cycle to achieve the intensity and overall duration of a normal cardiac rehabilitation program.

- Exercise therapy in PAD improves pain-free walking time by 189% and maximal walking time by 80% to 150% starting within 4 weeks of exercise, and benefit is maximized in 12 months. There is a 31% improvement in the ability to carry out ADLs and a 62% increase in physical activity.
- Mechanisms suggested to improve walking distance and functional abilities include angiogenesis, enhanced collateral blood flow from inactive to active muscles, improved blood and plasma viscosity, increased endothelial-dependent vasodilatation, and increased oxidative capacity.
- 🕮 Exercise improves PAD and walking economy by increasing biomechanical and metabolic efficiency.

LOWER EXTREMITY VASCULAR ULCERATIONS

Venous Stasis Ulceration

- **History and risk factors**: Venous HTN caused by valve damage and retrograde venous blood flow to superficial veins in chronic venous insufficiency, varicose veins, DVT, poor calf muscle function, arterio-venous fistula, sitting/standing for long periods, HTN, leg fractures/injuries, obesity, multiple pregnancies, previous surgeries (Frontera et al., 2020; Green, 2015)
- **Clinical findings**: Pitting edema is often present and worsens throughout the day. Hyperpigmentation of the skin is common secondary to hemosiderin deposits within macrophages, which stimulate melanin production. Chronic findings also include lipodermatosclerosis, which is characterized by induration and fibrosis of the dermis and subcutaneous layers lacking pitting edema. Venous eczema may also be presenting, which usually presents with erythema, scaling, pruritus, and weeping.
- **Location:** Venous ulcerations are most commonly located near the lower third of the leg, malleoli, and gaiter area and may be discrete or circumferential lesions.
- **Diagnostics:**
- Venous duplex ultrasound is the best method to test individual veins for venous insufficiency causing venous leg ulcers. CT or magnetic resonance (MR) venography can supplement this (Green, 2015; Moore et al., 2019).
- Continuous wave Doppler to test integrity of the venous system (Frontera et al., 2020)
  - Venous plethysmography to detect blood volume changes in an extremity including that of venous incompetence
- **Treatment**:
  - Compression is the mainstay of venous ulcer management.
    - Graded compression with the greatest pressure at the ankle (approximately 40 mmHg) tapering off to lower pressure below the knee (approximately 18 mmHg) increases limb hydrostatic pressure and reduces superficial venous pressure. Various compression bandage systems are available, including single and multilayer elastic bandage systems, short stretch bandages, and elastic tubular bandages.
    - Suggested approximately 14 to 17 mmHg pressure at ankle for compression stockings in mild varicose veins, approximately 18 to 24 mmHg for prevention of recurrence of venous ulcers on narrow legs and in slim patients for mild edema, and approximately 25 to 35 mmHg for chronic venous insufficiency and edema, as well as large heavy legs.
    - Compression with pneumatic devices has also been used.
    - Unna boot (a zinc oxide bandage) is commonly used; however, it risks the development of contact dermatitis.
  - Surgical debridement of nonviable tissue and shave therapy or excision of whole ulcers followed by skin grafting may expedite the healing process.
  - Other components of treatment: Leg elevation; exercise; autografts; topical agents like abx, growth factors, and fluid absorbents can help facilitate healing; debridement, usually with a curette, to stimulate intrinsic growth factors and wound closure; open or minimally invasive

techniques such as saphenous ablation and perforator ablation (Green, 2015; Mayrovitz et al., 2023; Montminy et al., 2018; Moore et al., 2019).

Arterial Ischemic Ulceration

- **History:** Intermittent claudication, rest pain, PAD, recent minor trauma, or localized pressure
- **Risk factors:** DM, smoking, obesity, heart disease, HTN, hyperlipidemia (HLD), progressive atherosclerosis (Green, 2015; Kirsner & Vivas, 2015; Sacchidanand et al., 2019)
- **Clinical findings:** Low amounts of exudates or edema compared with venous ulcers, pain even without signs of infection (pain may be present at rest and alleviated by hanging the foot over the side of the bed or sleeping in a chair; begins distal to the obstruction and moves proximal with disease progression), trophic changes, gangrene, absent or decreased distal pulses, possible bruits in more proximal arteries suggesting atherosclerosis, and delayed capillary refill time
- **Location:** Common locations include the toes, heel, ankle, dorsum of the foot, and bony prominences of the foot.
- **Diagnostics**:
  - ABI helps detect obstructive arterial disease via identification of decreased pressures in lower extremity vasculature
  - Transcutaneous O_2 values most helpful for evaluating severe ischemia; generally >55 mmHg considered normal, and wound healing is not reliable if <40 mmHg (Kirsner & Vivas, 2015; Sidawy & Perler, 2023)
  - Arterial duplex ultrasound evaluating arterial waveforms, pulse volume recordings (Kirsner & Vivas, 2015)
  - Arteriography, magnetic resonance angiography (MRA), and CT angiogram (CTA) to assess vascular anatomy and localize PAD in lower extremities (Sacchidanand et al., 2019)
  - Serum albumin, a marker for nutritional status: Lower levels linked to poor wound healing and associated with worse outcomes (Linn et al., 2020).
- **Treatment:** Goal is to increase peripheral blood flow to the affected area via percutaneous balloon angioplasty, laser angioplasty, or stent placement. Failure of the above or severe enough disease may necessitate amputation (Grey et al., 2006; Kirsner & Vivas, 2015; Mayrovitz et al., 2023).
  - ABI 0.5 to 0.3 with severe intermittent claudication and rest pain indicates severe arterial disease and requires urgent referral to a vascular specialist and possible arterial imaging.
  - ABI ≤0.3 with rest pain >2 weeks indicates critical ischemia and severe arterial disease with risk of limb loss requiring urgent referral to vascular emergency on-call team for possible intervention.

Neurotrophic Ulcers

- **History**: Painless but bleed with manipulation; chronic diabetes with neuropathy
- **Risk factors:** Diabetes, peripheral neuropathy, retinopathy, conditions leading to insensate lower extremity, which is then prone to cumulative microtrauma eventually developing into ulceration. Also age, reduced HDL, cholesterol, smoking, prior CVD (Adler et al., 2002; Green, 2015; Sacchidanand, 2019; Sidawy & Perler, 2023).
  - One study from the U.K. Prospective Diabetes Study in 2002 revealed that each 1% increase in HbA1c was associated with a 28% increased risk of PVD and each 10 mmHg increase in SBP was associated with a 25% increase in risk.
- **Clinical findings**: Punched-out lesions with a deep sinus, surrounded by inflammatory changes and callus
- **Location**: Typically occurs over pressure points or calluses (e.g., plantar surface of the first or fifth metatarsophalangeal (MTP) joint, base of distal phalanx of great toe, dorsum of interphalangeal joints of toes with flexion contractures, or callused posterior rim of heel pad)
- Chronic ulcers can be associated with arterial ischemia, venous stasis, and neuropathy, and the etiologies are not mutually exclusive.

- **Diagnostics** (Eastman & Dreyer, 2022):
  - Electrodiagnostics to identify any peripheral neuropathies
  - Vascular studies to differentiate from venous/arterial ulcers
  - Bloodwork/imaging to identify associated soft tissue/bone infections
- **Treatment:** Local wound care, management of secondary infections, debridement, pressure-offloading, proper footwear to distribute pressure, treatment of underlying disease (Sacchidanand et al., 2019)

MOST COMMON MAJOR PHYSICAL IMPAIRMENTS THAT OFTEN EXIST WITH CORONARY ARTERY DISEASE

Amputation

- The atherosclerotic vascular disease that affects the cardiovascular system also predisposes these patients to limb loss (dysvascular lower extremity amputation).
- There is a 75% coexistence of CVD, usually presenting as CHF or CAD, in persons undergoing dysvascular amputation. Long-term follow-up of dysvascular amputees shows that CHD is the most common cause of death.
- Diabetes, in addition to causing accelerated atherosclerotic vascular disease, is a major risk factor for amputation. It has been estimated that 50% to 70% of all amputations are the result of complications of diabetes.
- 🕮 Prosthetic ambulation is more energy-consuming than normal ambulation. The higher the level of amputation, the more energy is required per unit distance traveled. Amputees compensate by walking at slower speeds in order to keep the rate of energy expenditure stable (Table 9–10).
- For similar walking speeds, 25% more energy is required to walk with a below-the-knee prosthesis, 40% more for a bilateral below-the-knee prosthesis, 65% more for an above-the-knee prosthesis, and 100% more for bilateral above-the-knee prosthetic devices.
- 🕮 **The energy cost of ambulation for the amputee** is based on percentage increase above the cost of normal ambulation at 3 METs.

AMPUTEE EXERCISE STRESS TEST

- Upper extremity cycle ergometer stress test: First determine the safety and ability of mobility.
- Pharmacologic stress testing using dipyridamole for patients who are unable to perform any exercise

TABLE 9–10 **Energy Cost of Ambulation for the Amputee**

AMPUTATION	INCREASE IN ENERGY (%)	METS
No prosthesis with crutches	50	4.5
Unilateral BKA with prosthesis	9–28	3.3–3.8
Unilateral AKA with prosthesis	40–65	4.2–5.0
Bilateral BKA with prosthesis	41–100	4.2–6.0
Unilateral BKA plus contralateral AKA with prostheses	75	5.3
Bilateral AKA with prostheses	280	11.4
Unilateral hip disarticulation with prosthesis	82	5.5
Hemipelvectomy with prosthesis	125	6.75

AKA, above-the-knee amputation; BKA, below-the-knee amputation; METs, metabolic equivalents.
Source: From Flores AM, Zohman LR. Rehabilitation of the cardiac patient. In: DeLisa JA, Gans BM, eds. *Rehabilitation Medicine: Principles and Practice*. 3rd ed. Lippincott-Raven; 1998, with permission.

- Telemetry monitoring of ambulation training:
  - Preprosthetic period
  - Prosthetic period
  - Postprosthetic period

Stroke

- According to the medical literature, as many as 77% of stroke patients have some form of coexisting cardiac disease.
- The risk of stroke is doubled in individuals who have CAD.
- CAD accounts for the majority of subsequent deaths among stroke survivors (Stein & Brandstater, 2010).
- Acute cardiovascular events, such as an AMI or CABG, may be complicated by an acute stroke that occurs concomitantly.
- Roth et al. (1998) showed an overall incidence of cardiac complications of 27% to 34% during inpatient rehabilitation. The incidence was higher in patients with known CAD. Complications include:
  - HTN
  - Angina
  - MI
  - CHF
  - Cardiac arrhythmias

🕮 STROKE AND ATRIAL FIBRILLATION

- The prevalence of AFib is approximately 1% in the general population.
- It is 5.9% in those 65 years or older.
- AFib is present in 15% to 21% of the patients with stroke.
- Chronic, stable AFib increases the risk of stroke fivefold.
- Prevention of embolic stroke is best achieved by long-term anticoagulation with warfarin.
- 🕮 If risk of hemorrhage is high, aspirin 325 mg may be used as an alternative in patients with nonvalvular AFib.
- Aspirin is much less effective than warfarin in preventing embolism (Stein & Brandstater, 2010).
- 🕮 The 1996 Copenhagen Stroke Study measured the consequences of stroke with AFib compared with those with sinus rhythm:
  - Markedly poorer neurologic and functional outcomes
  - Higher mortality rates
  - Longer hospital stays
  - Lower discharge rates to home
  - Poorer outcome exclusively explained by initially more severe strokes
  - These results stress the importance of anticoagulant treatment of patients with AF (Jørgensen et al., 1996).

STROKE EXERCISE TESTING MODALITY

- Treadmill ambulation if it can be tolerated
- Stationary bicycle/leg ergometer modified for involved leg (ACE wrap)
- Portable leg ergometer that allows for seating in a wheelchair or arm chair
- Arm ergometer modified for involved hand or using one-handed arm ergometer
- Telemetry monitoring of level surface ambulation or general conditioning classes

HEMIPLEGIC AMBULATION COMPARED WITH NORMAL AMBULATION

- Speed: 40% to 45% slower
- Energy cost: 50% to 65% higher

Cardiovascular Conditioning of the Physically Impaired

CHOICE OF MODALITIES USED FOR ASSESSMENT DEPENDS ON NUMBER OF VARIABLES

- Upper extremity cycle ergometer: Impaired lower extremity with normal upper extremity
- Air dyne arm: Leg cycle ergometer for lower extremity weakness
- Hemiparetic: Strap the affected extremity to a foot pedal and/or handle bar
- Wheelchair-bound: Extra-wide treadmills that can accommodate a wheelchair

EVALUATION FOR RETURN TO EMPLOYMENT

1. Evaluation of the patient
2. Evaluation of the job
3. Matching the patient and the job
4. Other conditions

1. Evaluation of the Patient

- **Clinical evaluation to determine functional cardiac classification:**
  - New York Heart Association (NYHA) Class I: Can perform 7 METs or greater
  - NYHA Class II: Can perform 5 METs or greater but not 7 METs or greater
  - NYHA Class III: Can perform 2 METs or greater but not 5 METs or greater
  - NYHA Class IV: Cannot perform 2 METs or greater
- **Functional exercise stress test:**
  - Recommendations are made based on the maximum workload performance.
  - 7 METs or greater: Can return to work to most jobs in the United States
  - 5 to ≤7 METs: Can return to sedentary job and household chores
  - 3 to ≤4 METs or less: Not suitable to return to employment (Flores & Zohman, 1998)

2. Evaluation of the Job

- 🕮 **Physical task performed**
  The Department of Labor defines physical exertion requirements that are used in disability determinations (Social Security Administration, 2018):
  - Sedentary work: Lifting no more than 10 lb. Walking/standing occasional.
  - Light work: Lifting no more than 20 lb. Fair amount of walking/standing.
  - Medium work: Lifting no more than 50 lb. and frequent carrying up to 25 lb.
  - Heavy work: Lifting no more than 100 lb. and frequent carrying up to 50 lb.
  - Very heavy: Lifting more than 100 lb. and carrying more than 50 lb.
- **Environmental conditions at areas of work:**
  - Temperature and humidity: Hot and humid environment can increase the energy cost of work by two to three times.
  - Air pollution
  - High altitude
  - Motivation and emotional attitude of patients
  - Transportation to and from work
  - Household chores after work

3. Matching the Patient and the Job

- Matching the cardiac functional class and/or result of stress test to the requirement of the job
- Simulated job monitoring
- Monitoring the actual tasks at the job site

4. Other Conditions

- Emotional disorders
- Alcoholism
- Financial compensation (security gain)
- Retirement age
- Legal aspect
- Strenuous job requirements
- Patient motivation

AMERICAN HEART ASSOCIATION DIET

Step 1 Diet

- 8% to 10% of the day's total calories from saturated fat
- 30% or less of the day's total calories from fat
- Less than 300 mg of dietary cholesterol per day
- Just enough calories to achieve and maintain a healthy weight

Step 2 Diet

- Consider the Step 2 diet if you do not lower your cholesterol enough on a Step 1 diet or if you are at a high risk for heart disease or already have heart disease.
- <7% of the day's total calories from saturated fat
- 30% or less of the day's total calories from fat
- <200 mg of dietary cholesterol per day
- Just enough calories to achieve and maintain a healthy weight

BENEFITS DERIVED FROM LONG-TERM OUTPATIENT CARDIAC REHABILITATION

- Increased oxygen extraction and wider AVO_2 difference. Skeletal muscles take up more oxygen from the entering blood supply so that the venous return carries less back to the heart. The heart overall is doing less work to bring adequate oxygen to the tissue.
- Improved utilization of oxygen by active muscles resulting from increased oxidative enzymes and number of mitochondria in the muscles
- Increased maximal oxygen consumption ($\dot{V}O_2max$) or aerobic capacity and physical work capacity
- The conditioned patient generally has a slower pulse, a lower BP, and a lower RPP (RPP = HR × SBP). Because RPP is a good indicator of the myocardial oxygen demand ($M\dot{V}O_2$), trained cardiac patients function at a lower myocardial oxygen demand.
  - This results in a decreased $M\dot{V}O_2$ at rest and any submaximal workload.
  - Thus, an angina patient may be below the angina threshold in daily life and is able to perform certain activities without angina or silent ischemia.
- Increased CO at maximal exercise; CO remains the same at rest and at submaximal exercise:
  - CO = HR × SV
  - Fick equation: $VO_2max = (HR \times SV) \times A\dot{V}O_2$ difference
  - Therefore, $VO_2max = CO \times A\dot{V}O_2$ difference
- Increased SV at rest as well as submaximal and maximum levels of work. This increase is due mostly to a combination of increased blood volume and prolonged diastolic filling time (Flores & Zohman, 1993; Garden & Gillis, 1996).
- Exercise training, combined with intensive dietary intervention, with and without lipid-lowering drugs, results in regression or limitation of progression documented coronary atherosclerosis on angiography.
- Decreased myocardial ischemia as measured by exercise, EKG, and radionuclide perfusion imaging
- No apparent effect on development of a coronary collateral circulation and produces no consistent changes in cardiac hemodynamic measurement at cardiac catheterization

- Exercise training in patients with decompensated systolic HF resulted in documented improvement in functional capacity. The data reinforce that the favorable training effects in these patients are due predominantly to adaptation in the peripheral circulation and skeletal muscles rather than adaptation in the cardiac musculature (Cardiac Rehabilitation Guideline Panel, 1995).

CANCER REHABILITATION

GOALS OF REHABILITATION

The general rehabilitation goals of patients with cancer are similar to the general goals of patients with disabilities caused by other diseases. Rehabilitation of the patient with cancer should begin when disability is anticipated, rather than after it has occurred. The number of individuals surviving 5 years or more with a history of cancer continues to grow. Survivors may face significant physical and psychosocial problems that affect their quality of life. Rehabilitation goals can be appropriately assessed according to the different stages of the disease manifestation.

- Cancer rehabilitation is divided into four categories: Preventive, restorative, supportive, and palliative **(Table 9–11)**.
- **Cancer prehabilitation** is defined as a process on the continuum of care that starts from the time of cancer diagnosis to the beginning of acute treatment. Multimodal approaches include both physical and psychological assessments that establish a patient's baseline level of function and identify impairments. Once this assessment is complete, targeted interventions can be

TABLE 9–11 Classification of Cancer Rehabilitation

I. Preventive Rehabilitation – Begins after cancer diagnosis, even if no impairments of function – Done prior to or immediately after surgery, radiotherapy, or chemotherapy – The purpose of the rehabilitative measures at this point is to prevent impairments
II. Restorative Rehabilitation – The goal is the maximal recovery of function in patients with remaining function and ability
III. Supportive Rehabilitation – Augments self-care ability and mobility with effective methods (e.g., guidance with regard to self-help devices, self-care, and more skillful ways of doing things) for patients whose cancer has been progressing and whose impairments of function and abilities are declining – Additionally includes preventing the effects of disuse, such as contractures, muscle atrophy, loss of muscle strength, and decubitus ulcers
IV. Palliative Rehabilitation – While respecting patient wishes, allows patients with terminal diagnoses to live a high quality of life physically, psychologically, and socially – Aims to relieve symptoms (e.g., pain, dyspnea, and edema) and prevent contractures and decubitus ulcers using: ■ Heat ■ Low-frequency therapy ■ Positioning ■ Breathing assistance ■ Relaxation ■ Assistive devices

Source: Adapted from Shigemoto K, Abe K, Kaneko F, Okamura H. Assessment of degree of satisfaction of cancer patients and their families with rehabilitation and factors associated with it-results of a Japanese population. *Disabil Rehabil*. 2007;29(6):437–444.

applied to improve overall health and decrease the incidence and severity of impairments in the prehabilitation stage and beyond (Silver & Baima, 2013).

- **Preventive rehabilitation therapy** occurs before treatment, and may include education, exercises, and nutrition counseling, as well as address any current impairments.
- **Restorative therapy** focuses on maximizing functional recovery in patients with cancer who undergo treatment and have impairments.
- **Supportive rehabilitation therapy** is geared to those patients whose cancer is progressing and function is declining. The goals of supportive rehabilitation therapy include providing adaptive self-care equipment in an attempt to offset what can be a steady decline in a patient's functional skills. ROM and bed mobility can be taught to patients hospitalized or confined to bedrest to prevent the adverse effects of immobility, such as contractures, muscle atrophy, loss of muscle strength, and decubitus.
- In **palliative rehabilitation therapy,** the goal is to improve or maintain comfort and function during the terminal stage of the disease.
- At the end of life, the patient may have symptoms of pain, fatigue, and anorexia, leading to decreased mobility and independence. It is important to educate the patient and family about the body mechanics of transfer training, energy conservation, the use of assistive devices, and pain management in order for the patient to maintain a level of independence and quality of life (Table 9–12).
- Through retrospective analysis and observation, over half of all cancer patients had some form of difficulty addressable by physical medicine and rehabilitation. Physical medicine problems occur for patients with all tumor types.
  - In patients with central nervous system (CNS), breast, lung, head, and neck tumors, physical medicine problems may occur in >70%.
  - In one study of a group of 163 women with metastatic breast cancer, at least one physical impairment could be found in 92% of the group (Cheville et al., 2008).
  - There is often a gap in rehabilitative patient care that can dramatically be changed by introducing a physiatrist to the oncologic management team.
- While there is little literature studying the effectiveness of cancer rehabilitation, two studies have shown participation in a comprehensive inpatient rehabilitation resulted in significant

TABLE 9–12 Possible Contributions of Rehabilitation in the Various Phases of the Disease

TREATMENT
• Evaluating the treatment effects on function • Incorporating exercise, edema management, and increased activity to preserve and restore function • Managing pain with heat, cold, and TENS units
POSTTREATMENT
• Creating and supporting a program to assist the restoration of daily routines and promote a healthy lifestyle • Educating the patient about self-monitoring issues • Overseeing a maintenance program of exercise, edema management, and mobility
RECURRENCE
• Educating the patient and family regarding mobility training, appropriate body mechanics, and assistive devices
END OF LIFE
• Pain management (nonpharmacologic treatment) and symptom control • Maintaining independence and quality of life

TENS, transcutaneous electrical nerve stimulation.
Source: Adapted from Shigemoto K, Abe K, Kaneko F, Okamura H. Assessment of degree of satisfaction of cancer patients and their families with rehabilitation and factors associated with it-results of a Japanese population. *Disabil Rehabil*. 2007;29(6):437–444, with permission.

functional gains in patients who suffered from cancer or its treatment regardless of metastases or cancer type (Cole et al., 2000; Garden & Grabois, 1994). Functional Independence Measure (FIM) scores significantly improved in patients in all functional domains after receiving acute inpatient rehabilitation (Ng et al., 2017).

- Although cancer is not an explicit diagnosis in the Centers for Medicare & Medicaid Services (CMS) 60% rule of 13 compliant conditions to justify acute inpatient rehabilitation admission, several of its associated complications are, including:
  - SCI
  - Amputation
  - Brain injury
  - Fracture of hip/femur
  - Neurologic disorders (including polyneuropathy; Raj & Pugh 2019)

EPIDEMIOLOGY

- Approximately 1.9 million new cases are expected to be diagnosed in the United States in 2019. This estimate excludes basal and squamous cell skin carcinomas, which are not required to be reported to registries, and carcinoma in situ (noninvasive cancer) except for urinary bladder (American Cancer Society, 2023).
- The American Cancer Society reports that as of January 1, 2022, approximately 18 million Americans with a history of invasive cancer were alive. As of 2020, the cancer death rate for men and women combined had fallen 33% from its peak in 1991.
- The 5-year relative survival rate for all cancers diagnosed between 2012 and 2018 is 64% (American Cancer Society, 2023).
- A study of cancer patients referred to a 15-week multidisciplinary rehabilitation program resulted in clinically beneficial short- and long-term effects on health-related quality of life (van Weert et al., 2005).
- 🕮 The most common rehabilitation problems for the patient with cancer were described in a study by Lehman including a sample of 805 patients:
  - 35% General weakness
  - 30% ADL deficits
  - 30% Pain
  - 25% Difficulty with ambulation
- Other problems include speech, swallowing, respiratory, neurologic impairments, skin problems, nutritional deficiencies, lymphedema, skeletal disease, and psychological disorders (Lehman et al., 1978).

IMMOBILITY-RELATED PROBLEMS

- These complications are pertinent in the cancer patient secondary to longer time periods of illness, treatment, and recovery.

Immobility May Predispose Patients to the Following Body System Issues:

- Musculoskeletal:
  - Muscle atrophy
  - Decreased endurance
  - Joint contractures
- Vascular:
  - Orthostatic intolerance
  - DVT
  - PE

- Metabolic:
  - Protein loss
  - Impaired glucose tolerance
  - Hypercalcemia
- Skin: Pressure ulcers
- Gastrointestinal (GI)/genitourinary (GU):
  - Dysphagia
  - Difficulty voiding
  - Constipation
- Neuro: Compressive neuropathies
- Psychological:
  - Sleep disturbance
  - Depression
- Respiratory: Pneumonia

REHABILITATION INTERVENTION

- Traditional rehabilitation intervention may allow patients to maintain independence and AVOID complications associated with bedrest or immobility associated with prolonged hospitalization and medical treatment.
- Traditional rehabilitation intervention is geared toward countering the effects of bedrest and maximizing ADLs and functional mobility.
- Patients may perform strengthening exercises using an elastic band or mild resistive exercises against gravity while in bed.
- Bed mobility and frequent repositioning with cushions or pillows are to avoid skin breakdown and joint contractures:
  - Evaluation for an air mattress for those patients at high risk for skin breakdown
- Active range of motion (AROM) exercises are performed in the upper and lower extremity and are initiated whenever possible.
- A tilt table may be used to facilitate weight bearing and gradual upright positioning in those patients with orthostatic hypotension.
- Physical therapy (PT) and occupational therapy (OT) evaluations are necessary for equipment assessment, transfers, balance, ROM, bed mobility, and to maximize independence in ADLs and ambulation, as well as for patient and family education.
- 🕮 A high index of suspicion for swallowing disorders should be maintained.
  - Swallowing disorders may result from a variety of reasons in patients with cancer.
  - Swallowing difficulty can be associated with cognitive impairment, CNS involvement, and radiation treatment, and appear in patients with generalized deconditioning secondary to bedrest.
  - When aspiration is suspected, a dysphagia evaluation is performed. A follow-up videofluoroscopic swallow study (VFSS) may be required. If silent aspiration is suspected, VFSS is necessary.
- Calorie counts should also be performed to ensure adequate nutrition. A gastrostomy may be required for long-term nutritional management in some patients.
- 🕮 A high index of suspicion should be maintained for metastatic involvement of the extremities and spine.
  - Complaints of bony pain or discomfort warrant further investigation prior to initiating a skilled rehabilitation program.
  - Extremities with possible tumor involvement should be positioned for non-weight bearing and ROM withheld until a workup is completed.
- Patient-specific cardiac and pulmonary precautions should be ordered along with any skilled rehabilitation program (Lehman et al., 1978).

- Neurologic and muscular structures may be negatively impacted from the tumor invasion or side effects from cancer treatment.
  - For example, patients undergoing a modified radical neck dissection or selective neck dissection may suffer fewer impairments than someone undergoing a radical neck dissection. The sternocleidomastoid muscle, omohyoid muscles, internal jugular vein, and the spinal accessory nerve may be affected depending on the surgical procedure. Lymphedema after neck dissection also commonly occurs. Rehabilitation postoperatively, once safe to pursue, may include addressing lymphedema, posture and body mechanics, ROM, and strengthening of weakened muscles (Murphy et al., 2018).

CENTRAL NERVOUS SYSTEM TUMORS

Primary Brain Tumors

PRIMARY BRAIN TUMORS IN ADULTS

- Gliomas comprise approximately 60% of all primary CNS tumors.
- Cerebellar astrocytoma is the most common primary CNS solid tumor in young adults.

🕮 PRIMARY BRAIN TUMORS IN CHILDREN

- Brain tumors (26%) are only second to leukemia (28%) as the most prevalent malignancy in childhood (American Cancer Society, 2023):
  - Cerebellar astrocytoma is the most common posterior fossa tumor in childhood and has the best prognosis.
  - 🕮 Medulloblastoma is the next most common posterior fossa tumor in childhood and is the most prevalent brain tumor in children less than 7 years of age.
  - Brainstem gliomas are the third most frequent posterior fossa tumor in children.

BRAIN METASTASIS

- Brain metastasis occurs in approximately 25% of patients with cancer.
- In adults, >75% of cancers that metastasize to the brain originate from a primary tumor in the breast, lung, or melanoma.
- Lung cancer, followed by melanoma, accounts for most brain metastasis in men.
- Lung cancer, followed by breast tumors, accounts for most brain metastasis in women.
- Rehabilitation of patients with primary brain tumors or metastatic lesions is based on the location of the lesion and resultant neurologic and functional deficits (Takakura et al., 1982).

SIGNS AND SYMPTOMS OF BRAIN TUMORS

- Some of the presenting signs and symptoms of brain involvement are headaches, weakness, seizures, and cognitive impairment.
  - 🕮 Headache is the most common symptom.
  - 🕮 Weakness is the most common focal sign.
  - 🕮 Seizures are frequently the first presenting sign of CNS involvement.
- 🕮 Contrast MRI is the best diagnostic test.

MEDICAL TREATMENT OF BRAIN TUMORS

- Dexamethasone IV or orally to decrease brain edema and symptoms
- Antiepileptic medications to control or prevent seizures
- Neurosurgical evaluation to consider resection of single or a couple of metastases when indicated
- Whole brain radiation therapy (x-ray tomography [XRT])
- Stereotactic radiosurgery (radiation therapy), which gives high doses of radiation to a limited field, increases response rates to isolated metastasis and spares radiation injury to the rest of the brain.

- Chemotherapy and radiation may produce neurologic deficits, including impaired visual perceptual skills, memory, and judgment (Table 9–13).

CANCER REHABILITATION FOR BRAIN TUMORS

- Cognitive impairment, aphasia, dysarthria, and dysphagia require intervention through speech therapy, communicative evaluations, and dysphagia management. Deficits usually reflect the specific location of the lesion.
- Rehabilitation efforts are also geared toward the prevention of skin pressure breakdown through effective bed mobility, progressive ambulation, and mobilization, maximizing ADLs, safety, and equipment assessment, and family training to improve the patient's quality of life.

TABLE 9–13 Antineoplastic Agents Causing Neuropathy

AGENT	NEUROTOXICITY	COMMENT
Cytosine arabinoside	Acute cerebellar toxicity, encephalopathy, seizures, peripheral neuropathy	Rare
l-Asparaginase	Neuropsychiatric syndrome, cerebrovascular events	Rare
Methotrexate	Acute encephalopathy, leukoencephalopathy (with XRT)	Rare
Fluorouracil	Acute cerebellar dysfunction	(3%)
Fludarabine	Leukoencephalopathy	Rare
Oxaliplatinum	Distal sensory polyneuropathy, cold intolerance	Common (74% mild, 2% severe)
Cisplatinum	Peripheral sensory polyneuropathy, ototoxicity (20%), optic neuropathy (rare)	Common, dose-dependent, large sensory nerves
Carboplatinum	Peripheral sensory polyneuropathy, ototoxicity (5%–10%)	Less common/severe than Cis or oxaliplatinum
Paclitaxel (Taxol®)	Sensory/motor peripheral neuropathy (60% mild, 4% severe)	Common, dose-dependent, dose-limiting
Docetaxel (Taxotere®)	Sensory/motor peripheral neuropathy (49% mild, 3% severe)	Less common and less severe than Taxol
Vincristine	Peripheral sensory/motor polyneuropathy, autonomic neuropathy, cranial nerve palsy, encephalopathy	Almost 100%, dose-dependent
Vinorelbine	Sensory peripheral polyneuropathy	Common (25% mild, 1% severe)
Thalidomide, lenalidomide, pomalidomide	Sensory motor axonal polyneuropathy	Common, dose-dependent
Bortezomib (Velcade®), carfilzomib, ixazomib	Sensory peripheral polyneuropathy (36%), autonomic neuropathy (11%)	Very common, painful, dose-dependent/limiting
Ixabepilone	1%–24%	Reversible peripheral neuropathy
Immune checkpoint inhibitors	Immune-related, 2%	Myasthenia gravis, Guillain–Barré (acute and chronic demyelinating)

XRT, x-ray tomography.
Sources: Adapted from Abeloff M, Armitage J, Niederhuber J, Kastan M, McKenna W. *Clinical Oncology*. 3rd ed. Churchill Livingstone; 2004:1199–1205; McEvoy G, ed. *AHFS Drug Information 2008*. American Society of Health-System Pharmacists; 2008, with permission.

Spinal Cord Tumors

- Spinal cord tumors are rare.
- The majority of tumors affecting the spinal cord are extradural (95%) and arise from metastasis in the vertebral body.
- Approximately 70% of metastatic spinal tumors affect the thoracic cord.

RADIATION SIDE EFFECTS IN SPINAL CORD TUMOR TREATMENT

- Radiation therapy may also damage the spinal cord.
- Radiation myelopathy of the spinal cord is divided into transient myelopathy and delayed radiation myelopathy.

🕮 INDUCED TRANSIENT MYELOPATHY

- The most common form of radiation damage is **induced transient myelopathy**. The syndrome develops 3 to 4 months after treatment and spontaneously resolves over 3 to 6 months.
- There is transient demyelination of the posterior columns and lateral spinothalamic tract.
- Patients may report symmetric, electric shock-like paresthesias or numbness radiating to the extremities (Lhermitte's sign). CT scans are normal (Siker et al., 2016).

🕮 DELAYED RADIATION MYELOPATHY

- Delayed radiation myelopathy is irreversible. The symptoms do not begin until 6 to 12 months after completing radiation treatment. Most cases occur within 30 months.
- Symptoms begin with lower extremity paresthesias followed by bowel dysfunction and weakness. Midback pain may also be associated with radiation myelopathy.
- Resultant deficits depend on the level of neurologic involvement (Takakura et al., 1982).

Peripheral Nervous System Involvement in Cancer

- Peripheral neuropathy can occur due to tumor infiltration, paraneoplastic syndromes (NINDS website, 2019), or toxicity from the chemotherapy:
  - Peripheral polyneuropathy has been associated with lung cancer, multiple myeloma (MM), and breast and colon cancer.
  - Polyneuropathy is associated with inflammation and degeneration and can be demyelinating and axonal in nature.
- Symptoms include gait dysfunction, paresthesias, and sensory loss, as well as bowel and bladder dysfunction.
- Electromyography (EMG) may reveal fibrillation potentials and polyphasic motor unit potentials.
- Subacute motor neuropathy usually occurs with lymphoma. Anterior horn cells degenerate, resulting in weakness. However, stabilization occurs with gradual improvement.

CHEMOTHERAPY SIDE EFFECTS TO PERIPHERAL NERVOUS SYSTEM

- 🕮 Chemotherapy can cause a plexopathy or a peripheral polyneuropathy that is generally distal and symmetric. It is commonly associated with many agents (**Table 9–12**).
  - Vincristine may cause distal axonal degeneration, severe neuropathic pain, and in rare cases motor involvement, which can lead to quadriparesis.
  - Cisplatin, bortezomib, and vincristine may also cause autonomic neuropathies, resulting in fluctuating BP or HR.

RADIATION SIDE EFFECTS TO PERIPHERAL NERVOUS SYSTEM

- 🕮 Radiation may cause peripheral nerve damage due to effects on the nerve itself or by involvement of the surrounding connective tissue and vascular supply. Symptoms include muscle atrophy, hyperesthesia, paresthesias, decreased strength, and decreased ROM.

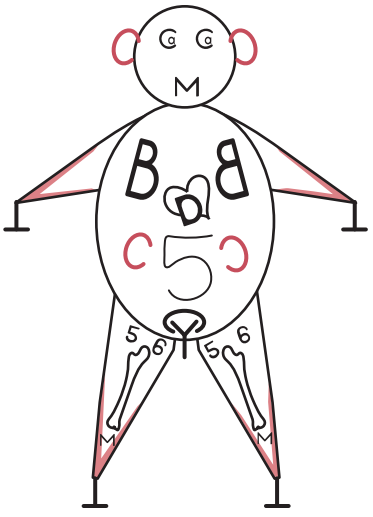
- 🕮 Brachial plexopathy is uncommon but can occur as a result of radiation treatment or through direct tumor extension, or paraneoplastic syndrome.
  - 🕮 Direct extension must be excluded especially in the presence of severe pain. In 90% of patients with direct tumor extension, pain is the initial symptom (Karandikar & Zakrasek, 2017).
  - 🕮 In radiation-induced plexopathy, numbness and paresthesias are typically the initial symptom. The *upper trunk* is predominantly involved with radiation plexopathy, and the *lower trunk* is predominantly involved in 75% of patients with invasive tumor.
  - 🕮 One example of tumor extension is Pancoast's syndrome, which is characterized by tumor invasion (bronchial carcinoma) into the superior pulmonary sulcus. It produces pain in the C8–T1 nerves (lower trunk plexopathy) as well as Horner's syndrome. Patients report pain beginning in the shoulder and vertebral border of the scapula. Radiation and surgery are the usual treatment (O'Young et al., 1997).
  - 🕮 Myokymia on EMG is pathognomonic of radiation-induced plexopathy. However, absence of myokymic discharges does not exclude radiation damage (Custodio, 2017).
- 🕮 MRI may reveal invasive lesions but is not 100% sensitive in detecting tumors causing a plexopathy. A CT reveals focal lesions in >90% of the cases.

TREATMENT OF CANCER-RELATED PERIPHERAL NEUROPATHY

- Treatment for involvement of the peripheral nervous system (PNS) usually focuses on treatment of the underlying malignancy, treatment of pain, or paresthesias.
- Supportive rehabilitation: Intervention including orthotics, assistive and adaptive equipment, endurance, energy conservation, ROM, skin protection, and maintenance of strength

OTHER CHEMOTHERAPY SIDE EFFECTS

CHEMOTHERAPY SIDE EFFECTS AND TOXICITIES ("CHEMO MAN")



C Cytarabine (conjunctivitis, keratitis)

M Methotrexate (myelosuppression, mucositis, liver fibrosis)

B Bleomycin + Busulfan (pulmonary fibrosis)

D Doxorubicin, daunorubicin (cardiotoxicity) + Trastuzumab (herceptin)

C Cisplatin + Carboplatin (ototoxicity/nephrotoxicity)

Y Cyclophosphamide + Ifosfamide (haemorrhagic cystitis)

5 Fluorouracil (myelosuppression, GI upset)

6 Mercaptopurine (myelosuppression)

■ Vincristine (peripheral neuropathy)

⊥ Taxanes e.g., paclitaxel (peripheral neuropathy)

GI, gastrointestinal.

- Cardiac:
  - Anthracycline agents, especially doxorubicin, can cause a range of cardiotoxic effects, from acute arrhythmias to mitochondrial damage, leading to congestive cardiomyopathy (Franklin, 2007).
- GU:
  - Hemorrhagic cystitis is a potential complication of chemotherapy treatment. It occurs most commonly with the use of ifosfamide or cyclophosphamide. Risk increases with IV use.

- Renal:
  - Patients receiving interleukin-2 infusions are monitored for hypotension and intravascular volume loss to prevent nephrotoxicity and prerenal azotemia. Interferon and bevacizumab can lead to proteinuria. Thus, patients receiving these treatments are screened appropriately (Franklin, 2007).

OTHER RADIATION THERAPY SIDE EFFECTS

- 🕮 Cognitive effects of radiation are probably dose-related. Young children are at higher risk than adults are, as myelin is developing rapidly; therefore, they are susceptible to CNS insult. It usually presents slowly, in delayed fashion, and can be difficult to distinguish from tumor recurrence.
- Cognitive effects are estimated in approximately 34% of patients after radiation therapy, especially if combined with chemotherapy.
- Fibrosis and contractures can develop. The patient should be maintained on a prophylactic stretching program before treatment and continued therapeutic stretching after radiation treatment.
- Postradiation osteonecrosis (avascular necrosis [AVN]) is uncommon but may lead to pathologic fractures.

PARANEOPLASTIC MYOPATHIES AND NEUROPATHIES

- These well-recognized syndromes can be associated with malignancies of the breast and lung.
- Rehabilitation treatment includes traditional rehabilitation, intervention, stretching, isometric exercises, assistive devices, energy conservation, and bracing, as well as social and vocational counseling. Specific attention must be paid to avoid exercise leading to fatigue.
- 🕮 **Carcinomatous myopathy** is a syndrome seen in metastatic disease that is consistent with muscle necrosis and presents with proximal muscle weakness.
- 🕮 **Carcinomatous neuropathy** affects peripheral nerves as well as muscle. Signs and symptoms include distal motor and sensory loss, proximal muscle weakness, and decreased reflexes and sensation. It most often occurs with lung cancer. Type II muscle atrophy is present as well as a distal peripheral polyneuropathy. Rehabilitative measures focus on supportive interventions including adaptive equipment, orthosis, and functional mobility.
- **Chemotherapy-related and steroid myopathies** result from atrophy of Type II muscle fibers in the proximal musculature.
- Isometric exercise may be used to improve muscle metabolism and enhance strength and recovery.

LYMPHEDEMA

- Lymphedema can occur as a result of direct tumor invasion, radiation, or surgery, or its treatment affecting lymph node drainage.
- Results from damage or blockage to the lymphatic system, resulting in accumulation of protein in the interstitium. This changes the colloidal pressure and detracts fluid into the interstitial space.
- 🕮 **Upper extremity lymphedema** is most common after breast cancer. Found in patients who have had a full nodal dissection, as well as radiation therapy to the axilla, and less commonly with sentinel node-directed biopsies.
- 🕮 **Lower extremity lymphedema** is associated with uterine disorders, prostate cancer, lymphoma, or melanoma:
  - Found in melanoma patients who had a nodal dissection
  - Prostate cancer patients with whole pelvic radiation or surgery

- Breast cancer patients experience upper extremity swelling and a sense of arm fullness. Usually develops over an extended period of time, postmastectomy or lumpectomy with full node dissection in up to one-third of women but only 5% after sentinel node biopsy.
- Patients with head and neck cancer may also experience lymphedema.

🕮 Lymphedema Presentation in Cancer

1. *Acute, transient, and mild:* Occurs a few days postoperatively
2. *Acute and painful:* Occurs 4 to 6 weeks postoperatively resulting from acute phlebitis or lymphangitis
3. *Erysipeloid form:* Results from minor trauma. Superimposed on chronic edema.
4. *Insidious and painless:* No erythema. Happens years after first treatment. Most common form.

Stages of Lymphedema

- Condition is described as progressing in stages:
  - **Stage 0** (latent): No edema. Damage to lymphatic vessels due to lymph node resection. Lymphedema is present but not clinically evident. Symptoms may include a sensation of heaviness of the limb.
  - **Stage 1** (spontaneously reversible): Edema usually upon awaking in the morning. Nonpitting edema. Limb size is normal or almost normal. Swelling may be reversible.
  - **Stage 2** (spontaneously irreversible): Pitting edema. Fibrosis starts to develop.
  - **Stage 3** (lymphostatic elephantiasis): Very large edema. Swelling is irreversible, and hard, fibrotic tissue develops.

🕮 Lymphedema Grading

- Lymphedema is categorized by its severity when compared with a healthy extremity:
  - **Grade 1** (Mild):
    - Pitting edema that can be reversed by elevation of the extremity. Present in distal arm or leg. Difference in circumference <4 cm.
  - **Grade 2** (Moderate):
    - Nonpitting, brawny edema not reversible with elevation of the extremity
    - The skin becomes hardened secondary to the development of fibrotic tissue due to chronic excess protein in the interstitial spaces and deposition of adipose tissue.
    - Edema moderate, reversible with effort. Involves entire limb or corresponding quadrant of the trunk.
    - Infection: None to occasional
    - Difference in circumference is >4 cm but <6 cm.
  - **Grade 3a** (Severe):
    - Swelling of the involved extremity described as lymphostatic elephantiasis. Edema is present in one limb and its associated trunk quadrant. Pressure does not produce any pitting. Edema minimally reversible or not reversible.
    - Cartilage-like
    - Infection common due to increased risk of skin breakdown. Usually fewer than four times per year.
    - The difference in circumference is >6 cm.
  - **Grade 3b** (Massive edema): Same symptoms as stage 3a except that two or more extremities are affected.
  - **Grade 4** (Gigantic): Elephantiasis:
    - Due to almost complete obstruction of the lymphatic channels
    - Edema is severe and irreversible. Impossible for ultrasound testing to even detect a pulse. Edema may involve face and head.
    - Infection frequent, more than four times per year
    - Focus is on management and containment of infections.

Lymphedema Diagnosis

- Diagnosis is based on the patient's history and a physical examination, as well as diagnostic procedures. Other causes of lymphedema or edema should be ruled out, including but not limited to malignancy, DVT, infections, and venous insufficiency.
- Limb circumference, water displacement, bioelectrical impedance, and optoelectronic perometry are used to measure limb differences.
- Lymphoscintigraphy is the imaging standard of care for identifying lymphatic system abnormalities. CT, MRI, and ultrasound have also been used to evaluate for lymphedema.
- Some genes have been identified that are associated with hereditary syndromes.

Lymphedema Treatment

- Without intervention, the limb enlargement can progress, which may result in significant social, physical, and psychological disability.
- Intervention focuses on prevention by not interfering with lymph outflow by constricting the arm, protecting the arm from infection and excess scarring, and avoidance of vasodilation from extreme heat exposure.
- Treatment involves elevation, retrograde massage or manual lymph drainage, or isometric external compression garment.
- **Elevation of the affected limb:** Reduces the amount of fluid and protein moving out of the capillaries by decreasing the hydrostatic pressure gradient from the vascular system to the tissues.
- Complete decongestive therapy is the standard of lymphedema therapy, and includes manual lymphatic drainage, compression bandages, appropriate exercise, and skin and nail care to attain a volume reduction, followed by maintenance of the limb size with a compression garment or bandaging.
- 🕮 **Compression therapy** by sequential graded pumps has been shown to be effective in reabsorption of water from the interstitium into the venous capillaries.
  - The downfall to this is that large protein molecules remain interstitial, continuing to change the colloidal pressure.
  - If a pneumatic or graded pump is used, it must be used for 2 to 6 hours daily followed by placement of a compression garment to prevent reaccumulation of fluid. This must be done daily for the remainder of the patient's life.
  - Precautions should be taken when using a compression pump, and its use should proceed after manual lymph drainage has been performed.
  - When initiating the compression pump, patients with cardiovascular compromise should be monitored closely for shortness of breath, increasing HR, fluctuation of BP, or complaints of increasing pain.
  - Caution should be taken in the presence of residual tumors.
  - Pumping should be discontinued if edema increases above the pump's sleeve.
  - Pumps should not be used in the presence of infection.
  - Pumps are contraindicated in bilateral mastectomy patients because truncal edema may result.
- 🕮 When more than one lymphedematous area is involved, there is no place for fluid resolution and other areas may become edematous.
- 🕮 Following mastectomy, immediate postoperative therapies can safely consist of hand pumping, hand and elbow ROM exercises, positioning techniques, postural exercises, and shoulder ROM exercises to 40 degrees of flexion and abduction. Active assistive exercises can be initiated when the surgical drains have been removed.
- 🕮 Additional management includes antibiotic treatment for sudden onset of warmth and pain suggestive of cellulitis or dermatolymphangitis. This helps to prevent exacerbation of the condition, as repeated infections damage the remaining lymphatic system in patients with known lymphedema. Bacterial antibiotic prophylaxis can be used in patients who suffer from recurrent cellulitis.

- **Corticosteroids** are helpful to decrease edema resulting from enlarged nodes.
- **Diuretics** are used if significant vascular impairment is a contributing factor to the development of edema. Only effective for short-term, acute management of lymphedema. For the chronic lymphedema patient, the diuretic effect is only temporary, and large protein molecules are left behind after its use.

METASTATIC BONE INVOLVEMENT (Coleman, 2001, 2006; Selvaggi and Scagliotti, 2005)

- In patients with advanced metastatic disease, the relative incidence of bone metastasis by cancer diagnosis is:
  - Breast cancer: 73%
  - Prostate cancer: 68%
  - Thyroid cancer: 42%
  - Lung cancer: 36%
  - Kidney cancer: 35%
- 25% have renal, thyroid, or some other type of primary cancer.
- In 25% to 30% of patients with bone metastasis, the bone lesion can be the initial presentation of the primary cancer (Morris et al., 2019).
- The presence of bone metastasis indicates advanced disease, and curative treatment may not be realistic by this stage (Morris et al., 2019). Once patients are found with metastatic bone disease, median survival is:
  - Breast cancer: 19 to 25 months
  - Prostate cancer: 12 to 53 months
  - Thyroid cancer: 48 months
  - Lung cancer: 6 to 7 months
  - Bladder cancer: 6 to 9 months
- 🕮 Bone is the most common site for metastasis and occurs in 70% of advanced breast, lung, kidney, thyroid, and prostate cancer (Coleman, 2006; Jehn, 2016). Skeletal metastasis arises through hematogenous spread.

🕮 Most Common Sites for Metastases

SEX	#1 SITE	#2 SITE	#3 SITE
Female	Liver	Bone	Lung
Male	Bone	Liver	Lung

Source: From Riihimaki M, Thomsen H, Sundquist K, et al. Clinical landscape of cancer metastases. *Cancer Med*. 2018;7(11):5534–5542. doi:10.1002/cam4.1697.

- 🕮 The most consistent symptom is pain that is most severe at night or upon weight bearing. In patients with spinal involvement, pain may be worse lying down and improves with sitting.
- Metastatic bone disease is the most common cause of cancer pain, and also causes pathologic fractures, neurologic injury, and functional disability. Pathologic fractures occur in 10% to 30% of patients with bone lesions (Morris et al., 2019)
- Skeletal-related events (SREs) are complications related to bone metastases and include pathologic fracture, spinal cord compression, and intractable pain requiring radiation therapy or surgery. SREs impact functional mobility, quality of life, psychiatric health, and overall survival (Jehn, 2016).
- 🕮 The risk of pathologic fracture is correlated with the extent of the lesion, the type of destruction, and the anatomic location. Lesions in higher stress areas such as the lesser trochanter are often associated with subsequent pathologic fracture. A high risk of fracture is associated with highly anaplastic and rapidly growing vascular lesions, which are usually osteolytic. The proximal femur is the most common site of pathologic fractures.

- 🕮 Skeletal metastases are rarely solitary. The axial skeleton is the most common site of bone metastasis, with other common sites including the femur, pelvis, humerus, ribs, and skull (Morris et al., 2019).
  - 70% of spinal metastases occur in the thoracic spine.
  - 95% are extradural in origin and involve the vertebral body anterior to the spinal canal.

CANCERS WITH A PREDILECTION TO METASTASIZE TO BONE	
Mnemonic	**Organ**
B	**Breast**
L	**Lung**
T	**Thyroid**
with a	
Kosher	**Kidney**
Pickle	**Prostate**

Diagnosis of Metastatic Bone Disease

- Some of the most prevalent cancers in the United States are commonly associated with metastatic bone disease. This is of particular clinical importance in breast and prostate cancers because of the prevalence of these diseases.
  - On postmortem examination, 70% of breast and prostate cancer patients had evidence of metastatic bone disease.
- However, bone metastases are not restricted to only these two cancers. They may complicate a wide range of other malignancies, resulting in considerable morbidity and complex demands on healthcare resources.
  - Carcinomas of the thyroid, kidney, and lung also commonly give rise to bone metastases, with an incidence at postmortem examination of 30% to 40%.
- If the clinician suspects metastatic involvement, the patient should be placed non-weight bearing on the involved extremity until a full evaluation and workup is completed.
- Complaints of pain warrant x-rays and potentially a bone scan and/or PET scans at any time during the disease.
- Bone scans and/or PET scans are typically done at the time of diagnosis with correlation of involved areas on the scan with x-rays. Most patients with high risk for conus medullaris involvement are followed with serial bone scans to detect bony involvement before any physical manifestations present.
- A **bone scan** is an important tool for detecting cancer that has metastasized. Bone scans can be falsely positive (light up as abnormal but not due to cancer) in the setting of prior trauma or degenerative joint disease, and many people have one or more of those conditions.
  - Bone scans usually pick up metastatic disease early. Bone scans are highly sensitive but not highly specific for tumor involvement. False negatives occur in the setting of bone destruction without ongoing repair or bone metabolism.
  - Up to one-third of patients with positive bone scans have no x-ray changes on plain films. If a cancer patient with normal spine radiographs complains of persistent back pain, a bone scan may be indicated. However, not all metastatic lesions are painful.
- A **PET/CT** scan with sodium fluoride (18F NaF) is a nuclear imaging test that scans the entire skeletal system and produces high-resolution images of the bones to detect areas of abnormal bone growth associated with tumors. They have become much more widely available.
  - PET scans are more sensitive (able to pick up disease better) and more specific (what they pick up is more likely to really be cancer) than bone scans.

- PET scans may be more specific and can detect bone metastasis when indicated for staging some malignancies (lung, breast, lymphoma, melanoma, esophagus, MM).
- A PET scan's high-resolution images and ability to scan the entire skeleton make it very helpful in detecting areas of abnormal bone growth associated with tumors.
- ^{18}F NaF PET scan imaging provides the physician with physiologic information of the bone. When the PET scan shows areas of increased uptake of ^{18}F NaF in the skeleton, it reflects sites of increased blood flow and bone remodeling. This information can be used by physicians to diagnose bone disease, detect bone injury, or determine the extent of metastatic disease.
- An additional type of PET scan is also utilized in clinical practice. It is fluorodeoxyglucose (FDG) PET scan and this scan shows areas of increased uptake of FDG in the skeleton. This reflects your body's use of radioactive glucose (FDG). This is because cancer cells take in glucose faster than normal tissue. Recent literature shows the ^{18}F NaF PET scan to be superior to FDG in identifying bony metastases (Araz et al., 2015; Zhang et al., 2018).

Bone Metastasis in the Upper Extremity

- 🕮 >90% of upper extremity metastases involve the humerus.
- In the upper extremity, the majority of symptomatic lesions are from:
  1. Breast cancer
  2. MM
  3. Renal cancer

Bone Metastasis in the Lower Extremity

- 🕮 Most metastases of the lower extremity involve the hip and femur.
- In the lower extremity, the majority of symptomatic lesions are from:

HIP	FEMUR
Prostate cancer	Breast cancer
Breast cancer	Renal cancer
Lung cancer	Multiple myeloma[a]
Lymphoma	Prostate cancer

[a]Multiple myeloma is a primary bone tumor that causes pathologic lesions.

🕮 *Note:* It is possible to have a negative bone scan with positive x-rays (any complaint of pain warrants a bone scan and plain film imaging).

Bone Metastasis in the Axial Skeleton

- Requires evaluation of the extent of metastatic involvement of the vertebral column. An MRI will clearly delineate spinal canal involvement even if plain radiographs are normal.
- 🕮 Denis (1984) described stability of thoracic and lumbar injuries by utilizing the three-column model described in **Figure 9–10**:
  - The spine is considered stable when only one column is involved unless it is the middle column.
  - The spine is considered unstable-when two or more columns are involved or the middle column is involved.
  - The spine is also considered unstable if >20 degrees of angulation is present.
  - These basic principles can be applied in evaluating metastatic bony involvement of the spine.

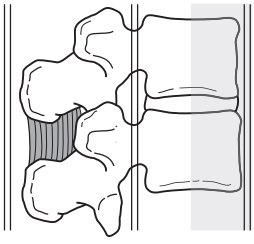
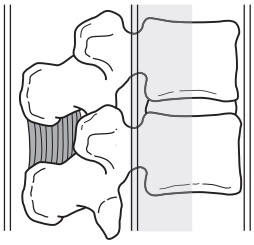
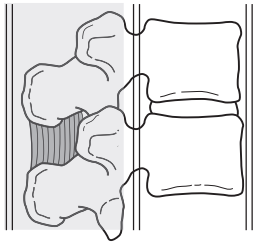
ANTERIOR COLUMN	MIDDLE COLUMN	POSTERIOR COLUMN
Anterior longitudinal ligament Anterior half of vertebral body Anterior annulus fibrosis	Posterior half of vertebral body Posterior annulus/posterior disc	Spinous process Laminae Facets
Anterior disc	Posterior longitudinal ligament	Pedicles Posterior ligamentous structures: Ligamentum flavum Intraspinous ligaments Supraspinous ligaments
 Anterior column A	 Middle column B	 Posterior column C

FIGURE 9–10 Three column model of spine stability. (A) Anterior column. (B) Middle column. (C) Posterior column.

- 🕮 The outcome for metastatic cancer to spine varies. It is better if the cancer is treated before spinal cord symptoms appear.
- Other prognostic indicators include the level of spinal cord involvement and the rate of neurologic progression. Breast cancer and lymphoma patients do better—they progress less rapidly and respond to multimodal therapy. Carcinomas of rapid progression that are less responsive to chemotherapy and radiotherapy do less well.

Treatment of Bone Metastases

- Once metastatic bone disease is identified, treatment may consist of radiation, chemotherapy, hormonal therapy, vertebroplasty, immobilization, splinting, bracing, bone resorption inhibitors (IV bisphosphonates or denosumab), and/or surgical intervention.
- Goals of treatment include pain relief, fracture prevention, functional maintenance or gain, and improved quality of life (Morris et al., 2019).
- If an unstable lesion is identified or suspected, a surgical opinion should be sought.
- Radiation therapy is efficacious in local tumor control and results in improved pain control in up to 85% of patients, with symptom relief occurring within the first 2 weeks.
- Radiation therapy promotes ossification of osteolytic bone metastasis and can reduce sensitivity of small pain fibers (Jehn, 2016).
- Palliative decompression and stabilization followed by radiation therapy can improve quality of life, even in aggressive or advanced disease (Jehn, 2016).
- 🕮 Admission to inpatient rehabilitation after surgical repair can help about half of patients who experience pathologic fracture become independent with ambulation.
- Poorer outcomes correlate with hypercalcemia and the need for parenteral narcotics (Franklin, 2007).
- Immobilization: Relieves pain and assists with prevention of pathologic fractures. Different types of immobilization can include:
  - Slings, splinting, and/or concomitant weight-bearing precautions with appropriate assistive devices
  - Cervical bracing: Halo bracing/cervical bracing, Philadelphia collars, sternal-occipital mandibular (SOMI) brace

- Other spinal bracing: Body jackets, such as plastic molded body jackets (thoracolumbosacral orthosis [TLSOs]), can be used for lesions involving the thoracolumbar region. A thoracic extension can connect a SOMI or Philadelphia collar to a custom-molded body jacket.

General Indications for Surgical Treatment of Metastatic Bone Disease

1. Intractable pain
2. Impending pathologic fracture
3. Pathologic fracture has occurred
4. Neurologic deficit (Jehn, 2016)

SURGICAL INTERVENTION IS INDICATED WHEN:

LOCATION	SIZE OF LESION	AMOUNT OF CORTEX INVOLVED
Upper extremity	>3 cm	>50%
Lower extremity	>2.5 cm	>30%–50%
Femoral neck (Figure 9-10A)	>1.3 cm	>1.3 cm in axial length
• Surgical intervention is indicated if >50%–60% of medullary cross-sectional diameter is involved. • This determination is enhanced by CT scan.		

Source: From Gerber LH, Vargo M. Rehabilitation for patients with cancer diagnoses. In: DeLisa JA, Gans BM. eds. *Rehabilitation Medicine: Principles and Practice*. 3rd ed. Lippincott-Raven; 1998:1293–1317.

- 🕮 Metastatic bone lesions can present as osteolytic or osteoblastic. Osteolytic lesions present net loss of bone by osteoclast-mediated bone reabsorption, while blastic lesions present as sclerotic areas of bone formation.
- Lytic lesions decrease the strength and stiffness of the bone, while blastic lesions decrease stiffness but do not change the strength of bone.
- 🕮 **Lytic lesions** (Figure 9–11) are generally considered to be more prone to fracture than blastic lesions. Lytic lesions typically occur in the following tumors:
  - Myeloma
  - Lung
  - Kidney
  - Thyroid
  - Malignant lymphomas
  - Breast
- 🕮 Median survival rates for these cancers *with* metastatic disease is:
  - Lung: 6 months. Aggressive course, metastases have higher risk of fracture. Cortical metastases are common in lung cancer.
  - Kidney: Variable, depends on medical condition. May be as short as 6 months.
- 🕮 Blastic lesions typically occur in prostate cancer (90%). The median survival rate for prostate cancer is 40 months.
- 🕮 Metastatic breast cancer can present as lytic or blastic bone lesions. Approximately 60% of breast cancer bone lesions are blastic. Median survival in breast cancer with only skeletal metastases is 24 months.

PRIMARY BONE TUMORS

- Metastatic cancer to the skeletal system is more common than primary bone tumors.
- Primary bone tumors, known as sarcomas, are very rare and account for 0.2% of all cancers in the United States each year.

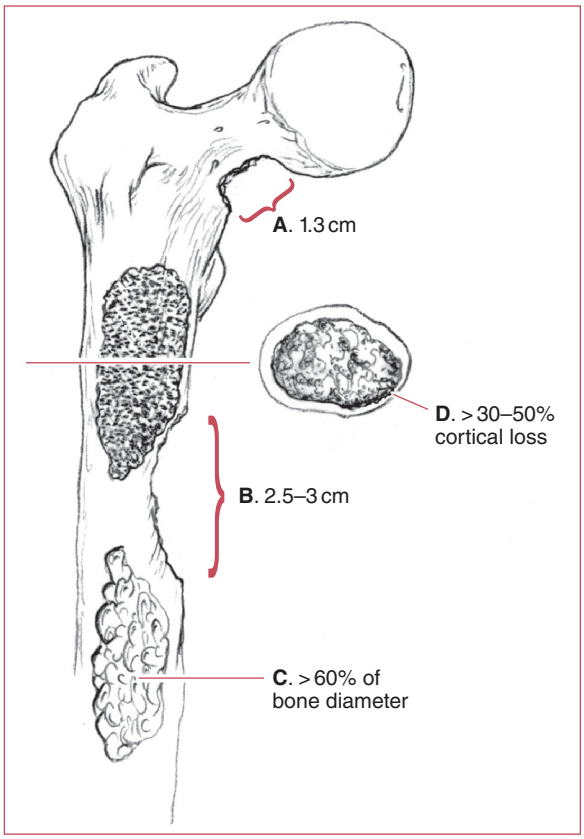


FIGURE 9–11 Lytic lesions of the femur that meet criteria for instability. (A) Cortical destruction exceeds 1.3 cm in the femoral neck. (B) Cortical destruction exceeds 2.5 to 3 cm elsewhere in the femur. (C) Lytic lesion exceeds 60% of the total bone width (diameter). (D) Lytic lesion exceeds 30% to 50% involvement of the cortex.

- Statistics about bone cancer (Key Statistics About Bone Cancer, 2019):
  - "Primary cancers of bones account for less than 0.2% of all cancers.
  - In adults, >40% of primary bone cancers are chondrosarcomas. This is followed by osteosarcomas (28%), chordomas (10%), Ewing tumors (8%), and malignant fibrous histiocytoma/fibrosarcomas (4%). The remainder of cases are several rare types of bone cancers.
  - In children and teenagers (those younger than 20 years), osteosarcoma (56%) and Ewing tumors (34%) are much more common than chondrosarcoma (6%)."

Osteosarcomas

- 🕮 The most common primary malignant bone tumor in children (Garden & Gillis, 1996)
- Occur in adolescence and commonly involve the knee and proximal humerus
- 5-year survival has increased to nearly 80% for localized osteosarcoma. For patients whose cancer has metastasized by the time the cancer is found, the 5-year survival rate is 15% to 30%. Survival is closer to 40% if only spread to lungs or if all of the tumor can be surgically resected (American Cancer Society, 2013).
- Treatment involves surgical intervention through amputation or limb salvage. Amputee and prosthetic management may be required (see Chapter 6, Prosthetics and Orthotics).

Multiple Myeloma

GENERAL

- While it is often considered a bony tumor, MM is actually a hematologic malignancy (plasma cell neoplasm) originating in the bone marrow.
- MM represents 10% to 25% of patients with pathologic fractures.
- Characterized by presence of cells resembling plasma cells originating in the bone marrow
- Occurs most commonly in patients 50 to 70 years old, males > females

- Usually presents with gradual onset of pain
- Frequently involves the lumbar spine, pelvis/sacrum, chest, skull, and ribs
- Often, there may be no early findings, and pathologic fracture may be the presenting manifestation of the disease.
- Course of disease is insidious and eventually leads to extensive marrow replacement, anemia, thrombocytopenia, and hemorrhages.

COMPLICATIONS

- Renal failure occurs as a result of tubular blockage by protein cast deposition.
- Bone involvement on radiographs reveals diffuse osteoporosis and multiple lytic lesions. However, early films are often negative.
- 🕮 Bone scans may be normal. However, a skeletal survey may reveal diffuse "punched out" lytic lesions with black sclerotic borders. PET scans can be helpful for staging.
- Amyloid deposits may also infiltrate peripheral nerves, causing a peripheral neuropathy.

TREATMENT

- Radiotherapy
- Chemotherapy
- Bisphosphonates
- High-dose steroids
- Intramedullary fixation may be difficult or impossible because of the remaining abnormal bone.
- Rehabilitation concerns are similar to those for patients with metastatic bone involvement compared with those with primary bone malignancies.
- A high index of suspicion is necessary to identify patients at risk for pathologic fractures.

REHABILITATION OF PATIENTS WITH ONCOLOGIC BONE DISEASE

- Goals are to protect the affected bone and promote strength and mobility.
- Crutches, walkers, wheelchairs, and required assistive devices and equipment are used to provide safety, joint protection, protected weight bearing, and function.
- Bracing (spinal orthosis) is indicated in patients with spinal instability. Corsets may be beneficial for pain relief and support when spinal stability is not a concern.
- Exercise programs should avoid high-impact, torsion, and manual resistive exercises.
  - Isometric and nonresistive isotonic exercises (swimming, walking, or stationary biking) are recommended within reason of each patient's current limitation. Exercises should improve endurance and strength.
- Fall prevention and proper body mechanics are essential.
- Physical modalities are used to relieve pain (soft tissue massage, transcutaneous electrical nerve stimulation [TENS] unit).
- Deep heat modalities such as ultrasound, diathermy, and microwave therapy are contraindicated in the presence of malignancy.

THE ROLE OF REHABILITATION IN PALLIATIVE CARE

- The goal of palliative care is to improve the quality of life of patients and their families who are facing serious illness through symptom alleviation, prevention, and relief of suffering.
- In January 2013, the U.S. District Court of Vermont clarified the Improvement Standard, which clarified Medicare guidelines to allow reimbursement of rehabilitation services to prevent or slow deterioration in the clinical condition.
- Rehabilitation goals are changed from return to prior level of function to address issues of mobility, independence, quality of life, and reduced burden of care (see **Table 9–12**).
- 🕮 Short, 5- to 7-day admissions to inpatient rehabilitation can benefit patients who choose to return home with hospice services by providing caregiver training in transfers, positioning, bowel and bladder management, and appropriate durable medical equipment use (Franklin, 2007).

  - Fall prevention should be emphasized to patients and their caregivers, particularly in the setting of metastatic bone disease.
  - In patients still undergoing outpatient treatments (e.g., radiation), timing of admission and discharge must be carefully considered to accommodate the duration of treatment.
- Medication regimens can be optimized in a safe and controlled manner in the inpatient setting to address pain, nausea, anorexia, constipation, mood symptoms, and altered mental status including agitation or delirium.
- Inpatient rehabilitation offers an opportunity to monitor and address a patient's nutritional status with ongoing speech therapy, laboratory testing, medication management, and appropriate nutritionist/dietitian consultation in a controlled setting.
- Data demonstrate that about half of cancer patients admitted to inpatient rehabilitation have below-normal prealbumin, which reflects compromised nutritional status.
  - Patients with poor nutritional status have prolonged hospital stays, more serious complications, and reduced quality of life (Franklin, 2007).
- Caretaker dependency, progressive debility, thoughts of uncontrolled pain and isolation, and loss of autonomy are the most distressing concerns.
- Quality of life is most dependent on physical strength, time spent in recumbency, and the ability to do what one wants.
- In patients with advanced disease, fatigue, pain, and generalized weakness are the most common reported symptoms.

Rehabilitation Goals and Interventions

- Assess patient care requirements and recommend environmental modifications upon discharge from hospital.
- Maintain patient's independence for as long as possible.
- Communicate with patient and family continually to reassess goals.
- Pain is a preventable symptom that affects 70% to 90% of patients with advance disease:
  - Physical modalities such as massage and cold can be used bedside to manage pain.
  - Cold packs should be applied to patient comfort.
  - Contraindications to cold: Insensate skin, atrophic skin, or skin exposed to radiation therapy.
- ROM and gentle strengthening can maintain strength and ROM.
- Prescription of assistive devices and instruction in compensatory strategies can aid in mobility.
- Environmental modifications to address the patient's functional decline can decrease the burden on the caregivers.
- Exercise can improve psychological status, combat deconditioning, and there is some evidence it may improve immune function.

TREATMENT OF CANCER PAIN

- Cancer pain may result from direct tumor invasion, chemotherapy, peripheral neuropathy, plexopathy, postprocedure pain, or can be unrelated to any of these factors. The World Health Organization (WHO, 1990) estimates that 25% of all cancer patients die with unrelieved pain.
- Pain can be effectively treated in 85% to 95% of patients with an integrated program of systemic, pharmacologic, and anticancer therapy.
- WHO has devised a three-step analgesic ladder to outline the use of nonopioid analgesics and adjuvant therapy for the treatment of progressively more severe pain (see **Figure 9–12**). See the following sections for a more detailed discussion.
- To maintain freedom from pain, analgesics should be given "by the clock," every 3 to 6 hours, instead of as needed or "on demand."
- For severe, chronic cancer-related pain, sustained-release opioids should be considered for increased compliance and consistent pain relief. Sustained-release oral opioids or transdermal patch can be added to the analgesic medication regimen.

Measuring and Assessing Pain

- Appropriate analgesic therapy is based on the pain level of the patient and the dosage of current medications.
- Pain can be measured on a scale of 0 to 10 (Wall & Melzack, 2000):
  - Pain levels of 1 to 4 are considered mild pain.
  - Pain levels of 5 to 6 are considered moderate pain.
  - Pain levels of 7 to 10 are considered severe pain.

🕮 World Health Organization Analgesic Ladder

- The three-step analgesic ladder developed by the WHO should be used to determine the appropriate level of analgesic therapy.
- *Step 1:*
  - Patients not on any analgesic therapy with mild/moderate pain are treated with Step 1 nonopioid analgesics (acetaminophen, ASA, nonsteroidal anti-inflammatory drugs [NSAIDs]).
  - Adjuvant pain medication may be added to facilitate better control or treat side effects or specific pain-related symptomatology. For example, amitriptyline has been shown to help with neuropathic pain and insomnia.
- *Step 2:*
  - If a patient has mild to moderate pain despite taking a nonopioid analgesic, the dose of the nonopioid analgesic should be maximized, and a Step 2 opioid analgesic ("weak opioid") should be added (codeine, hydrocodone, oxycodone).
  - Included in this step is tramadol, which is a centrally acting nonopioid analgesic with low affinity for mu-opioid receptors. Also inhibits reuptake of serotonin and norepinephrine. Max dose 400 mg/day.
  - Use adjuvant if needed.
- *Step 3:*
  - Patients who have moderate to severe pain despite therapy with Step 2 opioids require an increase in the dose of opioid or a change to Step 3 opioid when pain is severe (morphine, oxycodone, methadone, levorphanol, hydromorphone, fentanyl). Morphine is the agent of choice. Its dose should be maximized before other agents are added.
  - Again, use of adjuvant if needed
  - Patients who have mild to moderate pain while taking a Step 3 opioid should have the dose increased to an effective level (Levy, 1996).
  - Surgical intervention on the appropriate nerve may be indicated if pain relief is not completely effective.
- New adaptations of the WHO Analgesic Ladder have been proposed, which add a fourth step with advanced pain therapies for refractory pain or crises of chronic pain. Interventions include:
  - Parenteral opioids
  - Palliative treatment: Chemotherapy, radiation therapy, surgery
  - Nerve blocks
  - Spinal analgesics
  - Vertebroplasty
  - Radiofrequency ablation of tumor
  - Nerve abation
  - Neuromodulatio for increased compliance and consistent pain relief

Nonopioid Analgesics (Figure 9–11)

- Nonopioid analgesics are limited by maximum dosages.
- This group comprises anti-inflammatory agents (aspirin and NSAIDs) and acetaminophen.

Adjuvant Drugs (Figure 9–11)

- Adjuvant drugs include antidepressants, anticonvulsants, benzodiazepines, neuroleptics, antihistamines, corticosteroids, calcitonin, psychostimulants, and alpha-blockers. These supplement analgesics or treat side effects.
- Patients who do not respond to oral medications or have difficulty with limiting side effects may benefit from nerve blocks, TENS, or surgical intervention, such as cordotomy, dorsal column stimulation implantation (neuromodulation), or intrathecal administration of analgesics (opioid, local anesthetics, clonidine, or baclofen). Intractable coagulopathies may prevent the use of interventional pain procedures for pain management.
- Antidepressants including tricyclic antidepressants (TCA) and serotonin and norepinephrine reuptake inhibitors (SNRIs) can treat neuropathic pain disorders, which are commonly associated with cancer and its treatments. They can simultaneously address mood symptoms (Moryl et al., 2019).
- Anticonvulsants such as gabapentin and pregabalin effectively treat neuropathic pain but commonly cause drowsiness (DeLisa, 2010; Moryl et al., 2019).
- Tramadol is a centrally acting analgesic that mildly affects mu-opioid receptors and inhibits reuptake of serotonin and norepinephrine. It has been shown to effectively treat neuropathic pain and interacts with other serotonergic drugs such as antidepressants (Moryl et al., 2019).
- 🕮 NSAIDs are an effective adjuvant medication to reduce inflammatory pain, particularly when related to bone metastasis. They also may reduce bone resorption and bone tumor growth in osteosarcoma by inhibition of COX-2 (Tsuzuki, 2016).
- Their use is limited by side effects such as GI bleeding, renal toxicity, and effect on platelet function especially in patients with thrombocytopenia.
  - Misoprostol can serve as an effective protective regimen to reduce the risk of GI bleeding (Moryl et al., 2019).
- Topical treatments such as diclofenac gel and topical anesthetic patches or gels can provide localized analgesia if there is a desire for minimizing systemic effects.
- 🕮 Of the NSAIDS, ketorolac is the agent that has the least incidence of thrombocytopenia:

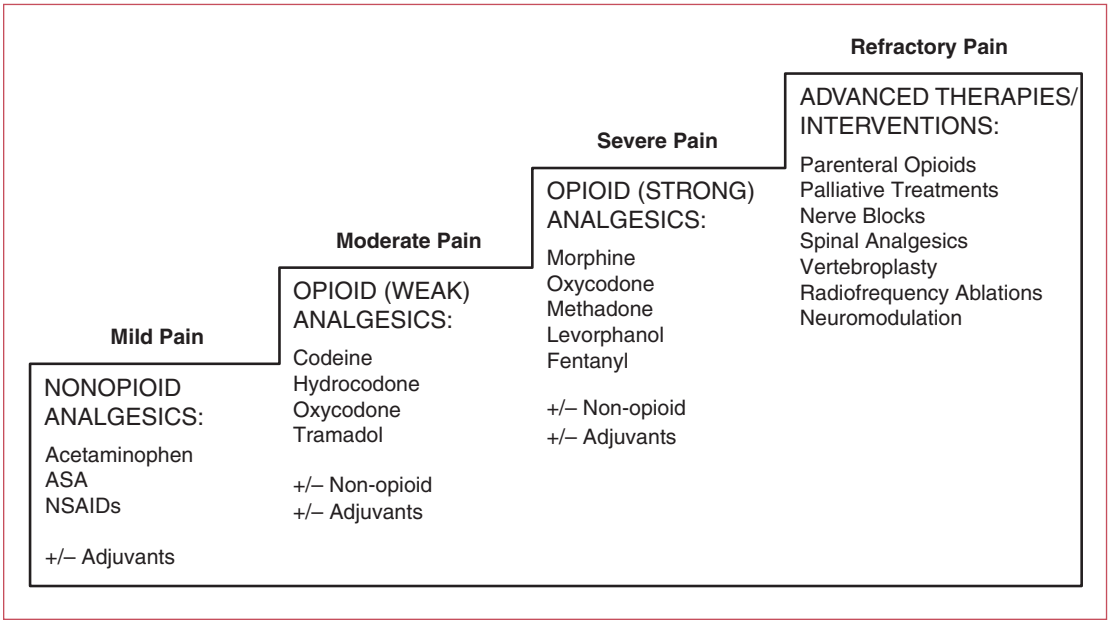


FIGURE 9–12 WHO Analgesic Ladder.
ASA, acetylsalicylic acid (aspirin); NSAIDs, nonsteroidal anti-inflammatory drugs; WHO, World Health Organization.
Source: Adapted from the WHO's cancer pain ladder for adults. World Health Organization. who.int/publications/i/item/9789241550390

  - As an alternative to narcotic analgesics, ketorolac is a potent COX-1 and COX-2 inhibitor and an antipyretic that can be used postoperatively.
  - It should only be used in the short term and with the lowest effective dose to prevent severe adverse effects such as GI bleeding, perforation, renal dysfunction, and reduced hemostasis.

Opioid Analgesics (Tables 9–14 and 9–15; Figure 9–11)

- Opioid analgesics have no ceiling, and dosing is guided by pain relief and is limited by side effects.
- Dosing should be titrated to a level in which pain is controlled or side effects limit increasing the dosage of the medication.
- Breakthrough pain is treated with "rescue doses." Use of short-acting, immediate-release preparations is recommended (oxycodone, morphine, hydromorphone). Calculated as 10% to 15% of the total daily opioid dose.
- While oral administration is the first choice, there are options for transdermal, rectal, IV, transmucosal, and spinal (epidural and intrathecal) routes.
  - These routes may be indicated for those patients for whom oral administration is not possible.
- An example of opioid analgesic agents and their conversion follows (**Tables 9–13** and **9–14**).

Treatment of Chronic Cancer Pain

- Adequate pain relief, appropriate rehabilitative programs and goals, as well as supportive psychosocial intervention continue to play an important role in improving the quality of life in patients with cancer.
- In order to treat chronic cancer pain, the physician should titrate around the clock medications and supplement with rescue doses for breakthrough pain. Rescue doses are based on one-sixth of the 24-hour total daily dose. Sustained-release medication should not be given on as-needed (PRN) basis.
  - Example: A patient taking 90 mg of controlled-release morphine every 12 hours would receive 30 mg of immediate-release morphine every 4 hours. The patient who is given q12 hour controlled-release morphine or oxycodone can appreciate analgesic effect in 1 hour, peaks in 2 to 3 hours, and lasts for 12 hours when the next scheduled dosing is due.
- Patients should be monitored closely, even daily, when beginning or changing an analgesic regimen. The optimal therapeutic regimen should be titrated based on unrelieved pain and side effects.
- It is important not to abruptly withdraw opioids. If pain has subsided, the dosage may be decreased 25% to 50% each day. If a patient has severe side effects from opioid use, one or two doses can be withheld and overall doses reduced by 50% to 75%.
  - Avoidance of abrupt discontinuation of opioids is essential to prevent physical withdrawal syndrome.
- Intrathecal drug delivery systems are an option when oral treatment does not provide adequate pain relief or when side effects are limiting activities of daily life. Doses can be varied and are lower compared with oral treatment, which may result in reduced side effects. Dose changes can be performed by a wireless computer.

MANAGEMENT OF GASTROINTESTINAL COMPLICATIONS

Nutrition

- The nutritional status of the cancer patient can become compromised as a result of radiation and chemotherapy.
- Radiation therapy or chemotherapy causes alteration in the saliva production and in taste. It may also cause mucositis, nausea, cramps, and diarrhea.
- Patients who receive radiation therapy may benefit from a lactose-free, low-residue oral diet.

TABLE 9–14 Opioid Analgesic Medications

OPIOID ANALGESIC	PARENTERAL (MG)	ORAL (MG)	CONVERSION FACTOR (IV TO PO)	DURATION IN HOURS (PARENTERAL, ORAL)
AGONISTS				
Morphine	10	30	3.0	3–4
Controlled-released morphine	–	30	–	12
MS Contin® Roxanol SR®	–	30	–	8
Methadone (Dolophine®)	10	20	2.0	4–8
Hydromorphone (Dilaudid®)	1.5	7.5	5.0	2–3
Fentanyl	100 mcg	–	–	1
Meperidine (Demerol®)	75	300	4.0	2–3
Levorphanol (Levo-Dromoran®)	2	4	2.0	3–6
Codeine	130	200	1.5	3–4
Oxycodone (Roxicodone®, component of Percodan®, Tylox®)	–	30	–	3–5
Hydrocodone (Lortab®, component of Vicodin®)	–	200	–	3–5
Propoxyphene (Darvon®, component of Darvocet®)	–	200	–	3–6
MIXED AGONIST-ANTAGONISTS				
Pentazocine (Talwin®)	60	180	3.0	2–4
Nalbuphine (Nubain®)	10	–	–	4–6
Butorphanol (Stadol®)	2	–	–	4–6

IV, intravenous; PO, orally.
Source: From Garden FH, Gillis TA. Principles of cancer rehabilitation. In: Braddom RL, ed. *Physical Medicine and Rehabilitation*. W. B. Saunders; 1996:1199–1214, with permission.

- When it is clinically appropriate to consider, when the patient is still on potentially active therapy, it is advisable to start parenteral nutrition when >20% of body weight has been lost.

Emesis

- Effective antiemetic management of the cancer patient includes the use of serotonin receptor antagonists such as ondansetron (Zofran), granisetron (Kytril), dolasetron (Anzemet), and palonosetron (Aloxi).
- Advantages of specific serotonin antagonists over conventional antiemetics such as metoclopramide include lack of extrapyramidal side effects, akathisia, and other CNS effects.
- Mild headache is, however, more common with these agents.

TABLE 9–15 Oral and Parenteral Dose Equivalents of Step 2 and Step 3 Opioid Analgesics

DRUG	DOSING	
	ORAL	*IV*
STEP 2 OPIOIDS		
Codeine	30 mg q4–6 hours	50 mg q4–6 hours
Dihydrocodeine	50–75 mg q4–6 hours	N/A
Hydrocodone	5 mg q4–6 hours	N/A
Oxycodone	5–10 mg q4–6 hours	N/A
STEP 3 OPIOIDS		
Morphine	15 mg q4–6 hours	5 mg q4–6 hours
Oxycodone	7.5–10 mg q4–6 hours	N/A
Hydromorphone	4 mg q4–6 hours	0.75–1.5 mg q4–6 hours
Fentanyl	200 mcg q4 hours (transmucosal); transdermal formulations and dosing also available	50 mcg/hour
NOT RECOMMENDED FOR ROUTINE USE		
Meperidine		50 mg q3–4 hours
Methadone		5 mg q6 hours
Levorphanol		1 mg q6–8 hours

IV, intravenous; N/A, not applicable.
Note: Oral opioids begin relief at 30 minutes and last approximately 4 hours. IV opioids begin relief at 5 minutes and last 1 to 2 hours.

MANAGEMENT OF FATIGUE AND DYSPNEA

- Etiologies of fatigue include cachexia, infection, anemia, and metabolic and endocrine disorders.
- Energy conservation techniques, work simplification, and assistive devices are used in the treatment of these patients.
- Frequent rest periods, pacing activities, easily accessible items, and decrease in upper extremity activities are all helpful.
- 70% of patients report fatigue chronically or while undergoing chemotherapy or radiation therapy.
- Metastatic disease to the lungs or pleural effusions can cause dyspnea.

BARRIERS IN CARE

- Certain patient populations can be difficult for therapists to gauge improvements with traditional measurements, as they are based on functional improvements. Patient populations include those with neurologic diseases such as ALS and advanced dementia, CVD such as CHF, COPD, and cancer.
  - ALS rehabilitation focuses on muscle atrophy, weakness, postural imbalance, spasticity, dysphagia issues, and gait disturbance.
  - Advanced dementia rehabilitation includes cognitive decline, loss mobility, and decreasing communication.
  - End-stage CHF and COPD rehabilitation includes combating fatigue and dyspnea.
- Research suggests healthcare professionals hesitate to discuss death and dying with their patients, thus delaying palliative care interventions.

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PEDIATRIC REHABILITATION

10

Roger Rossi, DO • Jessica Ace, MD • Michael Alexander, MD •
Michelle Sirak, MD • Sara J. Cuccurullo, MD

This chapter is designed to review various topics within the field of pediatric rehabilitation medicine that may be helpful when studying for the physical medicine and rehabilitation (PM&R) boards. It is broken down into different sections to encompass childhood development, growth, and the major childhood disabilities encountered in the field of rehabilitation medicine. The 11 subsections presented are:

1. Genetics and chromosomal abnormalities
2. Development and growth
3. Pediatric limb deficiencies
4. Diseases of the bones and joints
5. Connective tissue and joint disease
6. Pediatric burns
7. Pediatric cancers
8. Pediatric traumatic brain injury
9. Cerebral palsy
10. Spina bifida (myelodysplasia)
11. Neuromuscular diseases in children

GENETICS AND CHROMOSOMAL ABNORMALITIES

- Normal humans have 46 chromosomes (23 maternal and 23 paternal) in every cell except gonads, which have 23 chromosomes. Errors during cell division will result in chromosomal abnormalities.
- Chromosome abnormalities can be found in approximately 0.5% of all newborns.
- The numerical chromosome abnormalities are most frequently trisomy or monosomy.
- Chromosome abnormality should be suspected in children with any of the following:
  - Abnormal sexual characteristics
  - Congenital malformations
  - Developmental delay
  - Dysmorphic features
  - Intellectual disability
  - Prenatal and/or postnatal growth retardation
- Fragile X syndrome, XXY, and XYY often are associated with excessive growth.

PHENOTYPIC FEATURES OF SELECTED CHROMOSOMAL SYNDROMES (TABLE 10–1)

- Prenatal diagnosis includes amniocentesis at 14 to 16 weeks of pregnancy or chorionic villi sampling at 9 to 10 weeks of gestation.
- The exposure of a genetically susceptible fetus to a potential teratogen increases the chance of malformations. Although many environmental agents are potentially teratogenic, very few are proven teratogens. These include:
  - 🕮 Infectious agents, such as **TORCHES** infections (**T**oxoplasmosis, **O**ther agents such as Varicella virus or Parvovirus, **R**ubella virus, **C**ytomegalovirus, **H**erpes virus/HIV, **S**yphilis)
  - Drugs and medications (including alcohol, cocaine, anticonvulsants such as valproic acid, warfarin, vitamin A derivatives)
  - Maternal diseases (such as diabetes mellitus and phenylketonuria)
- Malformation can also be caused by uterine factors:
  - Malformed uterus
  - Twinning
  - Polyhydramnios

TABLE 10–1 Signs of Selected Chromosomal Syndromes

SYNDROMES	SIGNS
🕮 Trisomy 21 Down syndrome	Upward slant of palpebral fissures, Brushfield spots of iris, protruding tongue, third fontanelle, low-set auricles, excess nuchal skin, single palmar (simian) crease, single flexion crease and incurving (clinodactyly) of fifth fingers, increased distance between first and second toes, skin mottling, hypotonia, CHD such as endocardial cushion defect, VSD, and others
Trisomy 18 Edwards syndrome	IUGR, short palpebral fissures, small mouth, micrognathia, low-set abnormal auricles, prominent occiput, short sternum, abnormal position of fingers (second overlapping third and fifth overlapping fourth), hypoplastic fingernails, rocker-bottom feet, CHD, spasticity, feeding problems/failure to thrive
Trisomy 13 Patau's syndrome	IUGR, coloboma of iris (pupil of keyhole shape), capillary hemangioma, skin defect of skull, hyperconvex nails, polydactyly, rocker-bottom feet, arrhinencephaly, cleft lip and palate, CHD, urinary tract abnormalities
Prader–Willi syndrome (PWS) (chromosome 15 q11-13 – In 70% cases paternal chromosome deletion)	Infantile hypotonia, poor suck, hypogonadism, short stature, small hands and feet, obesity, cognitive impairment, behavioral problems
🕮 Turner's syndrome (45, X)	Short stature, triangular face, abnormal shape of ears, webbed neck, broad "shield" chest, wide-set nipples, congenital lymphedema of hands and feet, shortened fourth and fifth metacarpals and metatarsals, cubitus valgus, primary amenorrhea, CHD especially coarctation of aorta, mostly normal IQ, infertility
🕮 Klinefelter's syndrome (47, XXY)	Tall stature, postpubertally small testicles, gynecomastia, eunuchoid build, increased risk for mild ID, learning and behavior problems, infertility

CHD, congenital heart defects; IUGR, intrauterine growth retardation; ID, intellectual disability; VSD, ventricular septal defect.
Source: From Merenstein GB, Kaplan DW, Rosenberg AA, eds. *Handbook of Pediatrics*. 18th ed. Appleton & Lange; 1997, with permission.

INDICATIONS FOR GENETIC COUNSELING REFERRAL

1. Child with birth defects and/or developmental delay/intellectual disability
2. Dysmorphic child

3. Parent or sibling affected with known or suspected genetic disorder
4. Positive family history of birth defects or retardation in aunts, uncles, grandparents, or other relatives, especially if multiple members are affected
5. Possible teratogenic exposure or other abnormalities of pregnancy
6. Advanced maternal age (>35 years) or other indications for prenatal diagnosis

DEVELOPMENT AND GROWTH

- Development includes maturation of organs and systems; acquisition of physical, intellectual, and interpersonal skills; ability to adapt more readily to stress and assumption of personal responsibility; and capacity for creative expression. Growth signifies increase in size.

HEIGHT

- Birth length doubles by approximately age 4 years and triples by age 13 years.
- The average child grows approximately 10 in. (25 cm) in the first year of life, 5 in. (12.5 cm) in the second, 3 to 4 in. (7.5–10 cm) in the third, and approximately 2 to 3 in. (5–7.5 cm) per year thereafter until puberty.

WEIGHT

- The average infant weighs approximately 7 lb. 5 ounces (3.33 kg) at birth.
- Within the first few days of life, the newborn loses up to 10% of birth weight.
- Birth weight doubles between 4 and 5 months of age, triples by the end of the first year, and quadruples by the end of the child's second year. Between ages 2 and 9 years, the annual increment in weight averages about 5 lb. (2.25 kg) per year.

HEAD AND SKULL

- At birth, the head is approximately two-thirds to three-fourths of its total mature size, whereas the rest of the body is only one-fourth of its adult size.
- Six fontanelles (anterior, posterior, two sphenoid, and two mastoid) are usually present at birth.
- The anterior fontanelle normally closes between 10 and 14 months of age but may close by 3 months or remain open until 18 months.
- The posterior fontanelle usually closes by 4 months but in some children may not be palpable at birth.
- Cranial sutures do not ossify completely until later childhood.

OSSIFICATION CENTERS

- At birth, the average full-term infant has five ossification centers: Distal end of the femur, proximal end of the tibia, calcaneus, talus, and cuboid.
- The clavicle is the first bone to calcify in utero, beginning during the fifth fetal week.

BONE DEVELOPMENT

- 🕮 Greatest changes in bone mass occur in girls ages 12 to 15 years old and boys ages 14 to 17 years old.
- Rate of change in bone mass slows significantly from ages 16 to 18 in females and 17 to 20 in males, with peak bone mass being reached in both sexes between 25 and 35 years of age (Davies, 2005).

REFLEX DEVELOPMENT

- In neonates and infants, motor behavior is influenced by primitive reflexes as a result of the immature central nervous system (CNS).
- During the first 6 to 8 months of life as the CNS matures, these primitive reflexes are gradually integrated.
- Concurrently, more sophisticated postural responses emerge between 2 and 14 months that are incorporated into volitional motor behavior (Table 10–2).
- 🕮 Obligatory or persistent primitive reflexes are the earliest markers of abnormal neurologic maturation (see Table 10–2).

PHYSIOLOGIC POSTURAL REFLEX RESPONSES

TABLE 10–2 Postural Reflex Responses

POSTURAL REFLEX	STIMULUS	RESPONSE	AGE OF EMERGENCE	CLINICAL SIGNIFICANCE
Head righting	Visual and vestibular	Align face/head vertical, mouth horizontal	Prone: 2 months Supine: 3–4 months	Delays or absent in CNS immaturity or damage
Head and body righting	Tactile, vestibular proprioceptive	Align body parts in anatomic position relative to each other and gravity	4–6 months	Same as above
Protective extension tone or parachute reactions	Displacement of center of gravity outside supporting base in sitting, standing	Extension/abduction of lateral extremity toward displacement to prevent falling	Sitting anterior: 5–7 months; Lateral: 6–8 months; Posterior: 7–8 months; Standing: 12–14 months	Same as above
Equilibrium or tilting reactions	Displacement of center of gravity	Adjustment of tone and posture of trunk to maintain balance	Sitting: 6–8 months Standing: 12–14 months	Same as above

CNS, central nervous system.
Source: From Molnar GE, Alexander MA. *Pediatric Rehabilitation*. 3rd ed. Hanley & Belfus; 1999, with permission.

MILESTONES IN CHILD DEVELOPMENT

- Developmental milestones can be grouped into four distinct areas of function (Table 10–3):
  1. Gross motor behavior
  2. Fine motor, adaptive behavior
  3. Language behavior
  4. Personal/social behavior

AUTISM SPECTRUM DISORDER

- Autism spectrum disorder (ASD) is characterized by the *Diagnostic and Statistical Manual of Mental Disorders* (5th ed.; *DSM-5*; American Psychiatric Association, 2013) as encompassing disorders associated with both persistent deficits in social communication and interaction and repetitive, restricted patterns of interest, activity, or behavior.
  - 1 out of every 59 children are diagnosed with ASD, affecting males > females.
  - Deficits in social communication: Impaired ability to engage in communication, participate in social activities, and demonstrate nonverbal communication cues
  - Repetitive, restrictive behaviors, including hand flapping, finger flicking, echolalia, nonsense rhyming, repetitive questioning, and insistence on adherence to rules and routines

TABLE 10–3 Milestones in Child Development

AGE	GROSS MOTOR	FINE MOTOR ADAPTIVE	PERSONAL/SOCIAL	SPEECH AND LANGUAGE	COGNITIVE	EMOTIONAL
Newborn	Flexor tone predominates In prone, turns head to side Automatic reflex walking Rounded spine when held sitting	Hands fisted Grasp reflex State-dependent ability to fix and follow bright object	Habituation and some control of state	Cry State-dependent quieting and head turning to rattle or voice	Sensorimotor 0–24 months Reflex stage	Basic trust versus basic mistrust (first year) Normal symbiotic phase—does not differentiate self and mother
4 Months	Head midline Head held when pulled to sit In prone position, lifts head to 90 degrees and lifts chest slightly Turns to supine	Hands mostly open Midline hand play Crude palmar grasp Fascinated by own face in mirror	Recognizes bottle	Turns to voice and bell consistently Laughs, squeals Responsive vocalization Blows bubbles, "raspberries"	Circular reaction, the interesting result of an action motivates its repetition	Lap baby, developing a sense of basic trust
7 Months	Maintains sitting May lean on arms Rolls to prone Bears all weight, bounces when held erect Cervical lordosis	Intermediate grasp Transfers cube from hand to hand Bangs objects	Differentiates between familiar person and stranger Holds bottle Looks for dropped object Talks to mirror image	Uses single-word and double-consonant vowel combinations		At 5 months begins to differentiate mother and self—individualization Sense of belonging to a central person
10 Months	Creeps on all fours Pivots in sitting Stands momentarily, cruises Slight bowleg Increased lumbar lordosis, acute lumbosacral angulation	Pincer grasp, mature thumb to index grasp Bangs two cubes held in hands	Plays peek-a-boo Finger feeds Chews with rotary movement	Shouts for attention Imitates speech sounds Waves bye-bye Uses "mama" and "dada" with meaning Inhibits behavior to "no"	Can retrieve an object hidden from view	Practicing phase of separation—individuation, practices imitating separations

(*continued*)

TABLE 10–3 Milestones in Child Development (*continued*)

AGE	GROSS MOTOR	FINE MOTOR ADAPTIVE	PERSONAL/SOCIAL	SPEECH AND LANGUAGE	COGNITIVE	EMOTIONAL
14 Months	Walks alone, arms in high guard or midguard Wide base, excessive knee and hip flexion Foot contact on entire side Slight valgus of knees and feet Pelvic tilt and rotation	Piles two cubes Scribbles spontaneously Holds crayon full length in palm Casts objects	Uses spoon with overpronation and spilling Removes a garment	Uses single words Understands simple commands	Differentiates available behavior patterns for new ends, e.g., pulls rug on which is a toy	Rapprochement phase—individuation; ambivalence behavior to mother Stage of autonomy vs. shame and doubt Pleasures in control of muscle and sphincter
18 Months	Arms at low guard Mature supporting base and heel strike Seats self in chair Walks backward	Emerging hand dominance Crude release Holds crayon butt end in palm Dumps raisin from bottle spontaneously	Imitates housework Carries, hugs doll Drinks from cup neatly	Points to named body part Identifies one picture Says “no” Jargons	Capable of insight; problem-solving by mental combinations, not physical groping	
2 Years	Begins running Walks up and down stairs alone Jumps on both feet in place	Hand dominance is usual Builds eight-cube tower Aligns cubes horizontally Imitates vertical line Places pencil shaft between thumb and fingers Draws with arm and wrist action	Pulls on garment Uses spoon well Opens door turning knob Feeds doll with bottle or spoon Toilet training usually begun	Two-word phrases are common Uses verbs Refers to self by name Uses me, mine Follows simple directions	Preoperational period—2–7 years Able to evoke an object or event not present Object performance established Comprehends symbols	

TABLE 10–3 Milestones in Child Development (*continued*)

AGE	GROSS MOTOR	FINE MOTOR ADAPTIVE	PERSONAL/SOCIAL	SPEECH AND LANGUAGE	COGNITIVE	EMOTIONAL
3 Years	Runs well Pedals tricycle Broad jumps Walks up stairs alternating feet	Imitates three-cube bridge Copies circle Uses overhand throw with anteroposterior arm and motion Catches with extended arms hugging against body	Most children toilet trained day and night Pours from pitcher Unbutton; washes and dries hands and face Parallel play Can take turns Can be reasoned with	Three-word sentences are usual Uses future tense Asks who, what, where Follows prepositional commands Gives full name May stutter; eager Identifies sex of self Recognizes three colors	Preoperational period continues Capable of deferred limitation symbolic play, drawing of graphic images, mental images, verbal evocation of event	Stage of initiative vs. guilt 3–5 years Deals with issue of genital sexuality
4 Years	Walks down stairs alternating feet Hops on 1 foot Plantar arches developing Sits up from supine position without rotating	Handles a pencil by finger and wrist action, like adults Copies a cross Draws a froglike person with head and extremities Throws underhand Cuts with scissors	Cooperative play-sharing and interacting Imaginative make-believe play Dresses and undresses with supervision, distinguishing front and back of clothing and buttoning Does simple errands outside of home	Gives connected account of experience Asks why, when, how Uses past tense, adjectives, adverbs Knows opposite analogies Repeats four digits		
5 Years	Skips, tiptoes Balances 10 seconds on each foot	Hand dominance expected Draws man with head, body, extremities Throws with diagonal arm and body rotation Catches with hand	Creative play Competitive team play Uses fork for stabbing food Brushes teeth Self-sufficient in toileting Dresses without supervision except tying shoelaces	Fluent speech Misarticulation of some sounds may persist Gives name, age, address Defines concrete nouns—composition, classification, use Follows three-part commands Number concept to 10		Stage of industry vs. inferiority 5 years to adolescence Adjusts to the inorganic laws of the tool world

(*continued*)

TABLE 10–3 Milestones in Child Development (*continued*)

AGE	GROSS MOTOR	FINE MOTOR ADAPTIVE	PERSONAL/SOCIAL	SPEECH AND LANGUAGE	COGNITIVE	EMOTIONAL
6 Years	Rides bicycle Roller skates	Prints alphabet; letter reversals still acceptable Mature catch and throw of ball	Teacher is important authority Uses fork appropriately Uses knife for spreading Plays table games	Shows mastery of grammar Uses proper articulation		Stage of industry vs. inferiority continues
7 Years	Continuing refinement of skills		Eats with fork and knife Combs hair Is responsible for grooming		Period of concrete operational thought 7 years to adolescence Capable of logical thinking	

Source: From Molnar GE, Alexander MA. *Pediatric Rehabilitation*. 3rd ed. Hanley & Belfus; 1999, with permission.

- ASD may occur with or without intellectual disability and language impairment, as well as motor deficits including clumsiness, dyspraxia, gait dysfunction, and other abnormal motor signs including walking on tiptoes.
- Patients with ASD commonly have comorbid epilepsy.

PEDIATRIC LIMB DEFICIENCIES

CONGENITAL LIMB DEFICIENCY (TABLE 10–4)

- Congenital limb deficiencies (reduction defects) occur primarily during the first trimester when mesodermal formation of the limb occurs at day 26 of gestation and continues with differentiation until 8 weeks' gestation.
- 14/100,000 live births
- Risk factors for congenital limb deficiency include the drug thalidomide and maternal diabetes.
- 30% to 40 % reduced risk of limb deformity when taking folic acid during pregnancy
- There are three systems for classifying limb deficiencies:
  - International Society for Prosthetics and Orthotics (ISPO) classification system
  - Original (classic) classification
  - Frantz classification

TABLE 10–4 Examples of Common Deficiencies Named by Classification Systems

ORIGINAL (CLASSIC)	FRANTZ	ISPO
Upper extremity amelia	Terminal transverse	Transverse upper arm, total
Fibula hemimelia	Intercalary/normal foot Longitudinal/absent rays Fibular deficiency	Longitudinal fibular deficiency (total or partial)
Upper extremity phocomelia	Complete upper extremity phocomelia Distal/absent radius ulna Proximal/absent humerus	Longitudinal total, humerus, ulna, radius Carpal, metacarpal, phalangeal (total or partial)

ISPO, International Society for Prosthetics and Orthotics.
Source: From Molnar GE, Alexander MA. *Pediatric Rehabilitation*. 3rd ed. Hanley & Belfus; 1999, with permission.

International Society for Prosthetics and Orthotics Classification System

- 🕮 The ISPO classification is the preferred classification system.
- Classifies limb deficiencies as either transverse or longitudinal.
- Transverse deficiencies have no distal remaining portions, whereas longitudinal deficiencies have distal portions.
- The transverse level is named after the segment beyond which there is no skeletal portion.
- Longitudinal deficiencies name the bones that are affected:
  - Any bone not named is present and of normal form.

Original (Classic) Classification

- When describing limb deficiency, classic terms include the following:
  - Acheiria—missing hand or foot
  - Adactyly—absent metacarpal or metatarsal
  - Amelia—absence of a limb
  - Aphalangia—absent finger or toe

- Hemimelia—absence of half of a limb
- Meromelia—partial absence of a limb
- Phocomelia ("seal limb")—flipper-like extremity with an absent or markedly hypoplastic proximal limb and normal/nearly normal hand or foot

Frantz Classification

- Describes deficiencies as either terminal, representing the complete loss of the distal extremity; or intercalary, denoting the absence of intermediate parts with preserved proximal and distal parts of the limb.
- Those classifications are then divided into horizontal and longitudinal deficits.

CONGENITAL UPPER EXTREMITY DEFICIENCY

- Incidence is 1 in every 1,943 live births.
- Most cases of congenital upper extremity (UE) deficiency have no hereditary implications.
- Exceptions to the previously mentioned statement include:
  - Deficiencies that involve hands and feet
  - Central ray deficiencies
  - Adactyly involving the first four digits with the fifth intact
- 🕮 Craniofacial anomalies are associated with limb deficiencies.
- There are five associated syndromes seen with some limb deficiencies (Table 10–5).

TABLE 10–5 Associated Syndromes Seen With Limb Deficiencies

UPPER EXTREMITY SYNDROMES	ASSOCIATED PROBLEM
Thrombocytopenia-Absent Radius (TAR) Syndrome	Thrombocytopenia
Fanconi's Syndrome Anemia and leukopenia developing at 5–6 years of age	Anemia, leukopenia
Holt–Oram Syndrome Congenital heart disease, especially atrial septal defects and tetralogy of Fallot	Congenital heart disease
Baller–Gerold Syndrome Craniosynostosis	Craniosynostosis
VACTERL (or VATER) Syndrome Multiorgan symptom involvement	**V**ertebral defects **A**nal atresia **C**ardiac defects **T**racheo **E**sophageal fistula **R**enal dysplasia **L**imb deficiency

Source: Adapted from Molnar GE, Alexander MA. *Pediatric Rehabilitation*. 3rd ed. Hanley & Belfus; 1999, with permission.

Transradial Deficiency

- 🕮 The most common congenital limb deficiency is a left terminal transradial deficiency.
- 🕮 Prosthetic fitting should follow the attainment of normal developmental milestones, with the first fitting for a unilateral deficiency occurring when the child achieves sitting balance at around 6 to 7 months. The initial prosthesis has a passive mitt in which the infant can practice placing objects.
- 🕮 A more sophisticated prosthesis and terminal device is provided around 11 to 13 months when the child begins to walk, performs simple grasp and release activities, and has an attention span >5 minutes.
- The initial transradial prosthesis typically uses a self-suspending design with a supracondylar socket and a hand, which is preferred by parents.

- By age 4 to 5 years, the child can operate all types of prosthetic components and controls.
- 🕮 The **Krukenberg procedure** reconstructs the forearm and creates a sensate prehensile surface for children with absent hands by separating the ulna and the radius in the forearm:
  - Because of dramatic cosmetic appearance, the procedure is rarely used with unilateral conditions.
  - Indications include absent hands and visual impairment.

Transhumeral Deficiency

- For the transhumeral deficiency, the initial prosthesis may be suspended either by a harness or by silicone suction suspension.
- Prosthetic fitting again should generally follow the attainment of the normal developmental milestones. However, compared with fitting a child with a transradial deficiency, it is advisable to delay progression slightly to achieve optimal results. A transhumeral prosthesis can be more of an encumbrance than the transradial design to the infant, as the infant will have greater difficulty in rolling over with a transhumeral prosthesis.
- Active terminal devices should be prescribed shortly after the child begins to walk. The types of terminal devices used are the same as for the transradial:
  - Body-powered hooks are used successfully at 2 to 3 years old when the child is strong enough and has the cognitive ability to operate them.
  - By 18 to 36 months, a body-powered elbow may be used.
  - Externally powered devices may be used starting at 24 months.
- In general, the higher the limb absence, the less the child accepts the prosthesis, that is, transradial patients wear their prostheses more than transhumeral patients.

Upper Extremity Revision Amputation

- Required in 10% of UE congenital limb deficiencies.
- Treatment is directed at centralization of the hand and reconstructing the thumb.
- Examples:
  - Radial club hand
  - Ulnar club hand
  - These represent longitudinal deficiencies of the forearm.
- **Vilkke procedure:** Attaches a toe to the residual limb to create a pincer grip

CONGENITAL LOWER EXTREMITY DEFICIENCY

Fibula

- 🕮 Fibular longitudinal deficiency (fibular hemimelia) is the most common congenital lower limb deficiency. Prevalence of 1 to 2 per 100,000 live births. Bilateral deficiency occurs 25% of the time.
- Unilateral fibular deficiency creates a problem with limb length discrepancy. If leg length inequality is severe, a Syme's amputation may be performed with fitting of a Syme's prosthesis.

Tibia

- Transtibial deficiency (transverse deficiency of tibia) is more common than transfemoral (transverse deficiency of the femur).
- Longitudinal deficiency of the tibia occurs in one per one million births. Clinical picture includes a varus foot, a short leg, and an unstable knee, ankle, or both. Treatment of choice is a knee disarticulation.
- Partial tibial deficiency: 30% of partial tibial deficiency occurs as an autosomal dominant inherited pattern. Segment length of the tibia is important. If the tibial segment is long enough, the surgeon creates a synostosis with the intact fibula and amputation of the foot. This provides a stable walking surface for the child without a prosthesis.

Femur

- Partial proximal femoral focal deficiency (PFFD), also known as longitudinal deficiency of the femur, occurs in 1 per 50,000 births, and is the third most common longitudinal deficiency of the lower limb. It is usually unilateral (85%–90 % of cases), with bilateral involvement in 10% to 15 % of cases.
- PFPD is caused by impairment of proper development of the proximal femur and can cause stunting or shortening of the entire femur. The femur is typically short and held in flexion, abduction, and external rotation. There is proximal joint instability and inadequacy of the proximal musculature.
- Aiken's classification is most widely used.
- 🕮 70% to 80% of patients with PFFD also present with associated fibular deficiencies and cruciate ligament agenesis.

Treatment

- Orthotics (shoe lift) should be utilized in early development when the child begins to stand, in order to match leg length discrepancy and promote age-appropriate developmental milestones. Extension prostheses may also be utilized to add length (Modrcin et al., 2015).
- As the child gets older and the leg length discrepancy increases, surgery may be considered. One option is a limb lengthening procedure if the affected leg has adequate hip stability, and the remaining leg is functional with intact proximal and distal growth plates. Another option for PFPD is amputation of the foot on the affected side (with a Syme or Boyd amputation), with fusion of the shortened femur to the tibia, leaving a residual limb that will accept an appropriate above-the-knee prosthetic with a mechanical knee (Modrcin et al., 2015).

Options

- Van Ness Rotation: Controversial procedure that allows simulation of below-knee function by rotating the foot by 180 degrees so ankle motion can control the prosthesis.
- Nonstandard prosthesis or shoe lifts with no surgical conversion.

Fitting Timetable of Lower Extremity Amputee

- The lower limb-deficient child is fitted with a prosthesis when ready to pull up to standing position at 9 to 12 months. It is advisable to fit a jointless, above-the-knee prosthesis to the toddler.
- The normal child does not establish heel-to-toe gait until around 2 years.
- Prosthetic heel strike to toe-off gait is not attained until 5 years or when the child can demonstrate sustained one-legged standing.
- A knee joint is usually added as early as 18 months of age.

Components

- 🕮 The most common prosthetic foot prescribed for the child amputee has been the solid ankle cushion heel (SACH) foot, although energy-storing feet for children are becoming more popular.
- Some knees that can be used in children include the following:
  - *Single axis knees* (with or without locks) are durable and lightweight.
  - *Polycentric knees* are good in situations in which the residual limb is long and the knee centers are difficult to match.
  - *Fluid-controlled knees* offer a smoother gait and the ability of the knee to adapt to different walking speeds. Fluid-controlled knees are reserved until adolescence secondary to size and weight restraints.

- Suspension systems should be easily adjustable to allow for growth. Suspension sleeves and silicone suction suspensions provide good adjustability because they allow for growth and provide excellent suspension. A suction socket is not prescribed until a child can assist in donning a prosthesis, at about 5 years of age. The pelvic belt is an acceptable way to suspend an above-the-knee prosthesis.
- Below-the-knee amputees may use a patellar tendon-bearing prosthesis with a supracondylar cuff. It should be noted that one-third of limb-deficient children using this type of suspension develop a dislocated patella.
- Major causes of gait deviations are growth or worn prosthetic parts.
- Prostheses need to be replaced more frequently in the growing child.

AGE	CHILD QUALIFIES FOR:
Until 5 years old	New prosthesis yearly
Between ages 5 and 12 years old	New prosthesis every 18 months
Between ages 12 and 21 years old	New prosthesis every 2 years

- Children may assume gait deviations to relieve pressure if they have an ill-fitting prosthesis.

ACQUIRED AMPUTATIONS

Causes

- The most common cause of pediatric acquired amputation is trauma, occurring two times more frequently than disease-related amputation:
  - Motor vehicle, motorcycle, and train accidents account for the majority of childhood acquired amputations.
  - Home accidents (e.g., burns, fireworks) are also a common source of acquired amputation in the pediatric population.
- Single limb loss occurs in more than 90%, with 60% involving the lower extremity (LE). Boys are affected greater than girls with a ratio of 3:1.
- The most frequent cause of disease-related amputation is childhood tumors, with the highest incidence of malignancy occurring in the 12 to 21 age group. Osteogenic sarcoma and Ewing's sarcoma occur most commonly (please also see Pediatric Cancers section). Other disease-related causes include:
  - Vascular insufficiency—gangrene
  - Neurologic disorders—for example, neurofibromatosis with associated nonunion of fracture
  - Emboli from meningococcemia may cause autoamputation of limbs or digits and can affect all four limbs.

Prosthetic Considerations

- The management of an acquired amputation is different in children. It is important to retain the bony growth centers at the distal femur as well as the proximal and distal tibia to allow for continued longitudinal growth of the residual limb.
- By performing a joint disarticulation (rather than a transmetaphyseal or transdiaphyseal amputation) in a growing child, the epiphyseal growth plate can be preserved:
  - In the femur, for example, 70% of growth occurs from the distal physes. The loss of the distal femoral epiphyses can result in a significantly shorter residual limb.
- Because of the natural tendencies of transected immature bone, terminal bony overgrowth can also occur. A knee disarticulation preserves the distal femoral epiphysis, ensuring continued growth and avoiding distal bony overgrowth. It also provides a weight-bearing distal end and a long femoral lever arm for enhanced suspension and decreased energy expenditure with ambulation:

  - A disadvantage of knee disarticulation may be less ideal cosmesis and fewer prosthetic knee options.
- As the child grows, the limb with the amputation will not grow as large, allowing for cosmetic fix. If the epiphyseal growth plates are damaged during surgery, expandable prostheses at the proximal and distal ends of the femur and the proximal end of the tibia allow periodic lengthening of the device.

Complications

- 🕮 Terminal overgrowth at the transected end of a long bone is the most common complication after amputation in the immature child, occurring most frequently on the humerus, fibula, tibia, and femur, in that order. The appositional growth may be so significant that the bone can pierce the skin and cause ulcers:
  - Treatment of choice is surgical revision.
- Other complications include bone spur formation, development of adventitious bursae, and stump scarring requiring socket modifications.
- Fitting the child with an acquired amputation follows the same timetable as a congenital amputee except for the fact that a child who undergoes an amputation will require a temporary or preparatory prosthesis while postoperative swelling subsides.
- Intraoperative prosthetic fitting for an LE amputee may be performed:
  - *Advantages:*
    - Allows amputee to begin walking soon after surgery
    - Decreased edema and calf pain
    - Good candidates include teenagers or young adults undergoing amputation for a tumor.
  - *Disadvantages:*
    - Weight-bearing restrictions and activity restrictions may put the stump at risk.
    - Poor candidates for this procedure include young children who do not understand the restriction, immunocompromised children, and children with insensate limbs or infections.

GENERAL FUNCTIONAL ISSUES

- The child with isolated limb deficiency or amputation is capable of achieving age-level academic skills.
- Amputees preserve their energy expenditure by decreasing their walking speed.
- 🕮 Motorized wheelchairs are traditionally introduced when a child is 5 to 6 years old. In exceptional cases and with innovative technology, children as young as 1 to 3 years old have been reported to use power mobility consistent with developmentally appropriate mobility expectations. Cognitive skills of spatial relation and problem-solving are essential predictors of power mobility (Alexander and Matthews, 2015; Tefft et al., 1999).
- The child with bilateral UE deficiencies will attempt to use their feet for fine motor tasks and should be encouraged to do so at an early age.

PHANTOM PAIN

- 🕮 Children with congenital limb deficiency do not develop phantom sensation or pain even after conversion to surgical amputation of the limb.
- However, children with acquired amputations retain some awareness of the amputated part. This sensation has been described as uncomfortable or painful.
- The older the child is at the time of the amputation, the greater the chance that they may experience phantom pain, especially if the amputation occurs after the age of 10.

DISEASES OF THE BONES AND JOINTS

THE FEET AND TOES

Metatarsus Varus (Figure 10–1)

- Characterized by adduction of the forefoot on the hindfoot, with the heel in normal position or slightly valgus.
- Flexible deformities are secondary to intrauterine posture and usually resolve.
- Rigid deformities may require splinting.
- 85% correct by age 3 to 4 years.

FIGURE 10–1 Metatarsus varus.

Club Foot (Talipes Equinovarus)

- Characterized by a congenital malalignment of the talocalcaneonavicular complex.
- Incidence is 1 per 1,000. Club foot follows a hereditary pattern and may be part of a generalized syndrome or be associated with anomalies, especially of the spine.
- Club foot presents with the following deformities on physical exam, remembered by the mnemonic **CAVE**:
  1. Midfoot **C**avus
  2. Forefoot **A**dductus
  3. Hindfoot **V**arus
  4. Hindfoot **E**quinus
- Patients will also exhibit calf atrophy and leg length discrepancy.
- Diagnosis is clinical and includes grading each major component of club foot in order to assess overall severity. The most widely used grading systems are the Pirani and Dimeglio scores. Foot bimalleolar (FBM) angle can also be assessed to score and monitor the progression of club foot.
- 🕮 Conservative treatments are taken first, with the Ponseti method of serial casting and manipulation considered first-line treatment in the United States. Success of this therapy is highest when it is initiated in the first month of life. A recent study showed a low correlation between Pirani and Dimeglio scores with the number of casts required for the correction of club foot. There was no correlation between FBM angle and casting.
- Less frequently, the French functional physical therapy (PT) method is also used, which employs daily manipulation of the foot in combination with temporary post-manipulation immobilization by bracing and splinting. Other PT programs and bracing methods may also be helpful. Surgical correction is the last line of treatment and is used in resistant, persistent, or relapse of deformity.

Talipes Calcaneovalgus

- Excessive dorsiflexion at the ankle and eversion of the foot.
- Usually due to intrauterine position.
- Treatment includes stretching and rarely splinting.

Flat Foot (Pes Planus)

- Normal condition in infants.
- Prevalence as high as 80% in children.

Cavus Foot

- Unusually high longitudinal arch.
- May be hereditary or associated with neurologic conditions, such as poliomyelitis, Charcot–Marie–Tooth (CMT) disease, or Friedreich's ataxia.
- Usually associated contracture of toe extensors (claw toes).

Claw Toes

- Metatarsophalangeal joints are hyperextended and interphalangeal joints flexed.
- Usually congenital and seen in disorders of motor weakness, such as CMT or pes cavus foot deformity.

THE LEG

Genu Varum (Bowleg; Figure 10–2)

- Infants generally have bowing of the legs as a normal finding.
- By 12 to 18 months of age, the legs have straightened and progressed to mild knock-knee (genu valgus).
- They then gradually assume their ultimate configuration by 6 to 7 years of age.

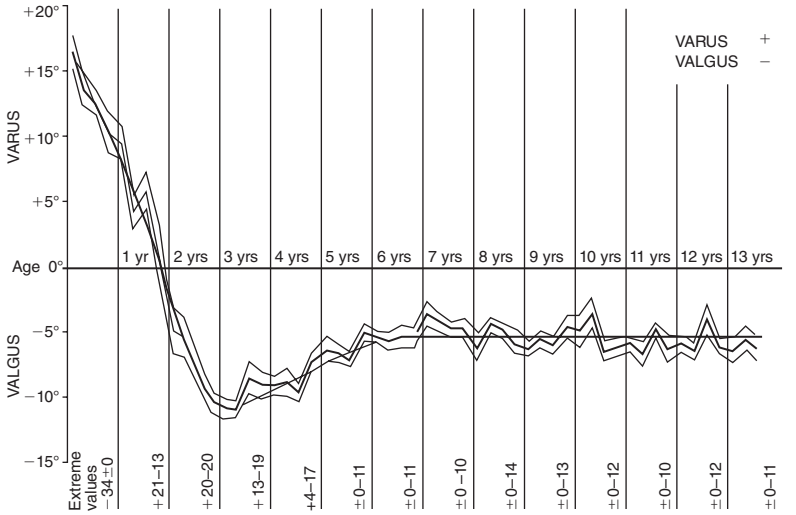


FIGURE 10–2 Genu varum (bowleg).

Blount's Disease (Tibia Vara; Figure 10–3)

- Due to abnormal function of the medial portion of the proximal tibial growth plate and results in bowing in the proximal tibia.
- It is the most common morphologic cause of bowing in the young child and is found most commonly in obese children who walk at 9 to 10 months.
- It is more common in African Americans than other racial groups and should be suspected in all children with persistent bowing after 2 years of age.
- Treatment is usually osteotomy of the proximal tibia and fibula, which may have to be repeated one or more times.

THE HIP

Developmental Dysplasia of the Hip

- Preferred term for what was previously known as congenital dislocation of the hip. It includes hip subluxation, hip dislocation, and acetabular dysplasia, all of which imply instability of the hip.
- Hip dislocation is usually diagnosed at birth, but acetabular dysplasia may present several months later.
- Hip dislocation occurs in around 1 per 1,000 births; more common in breech babies and females than in males.
- If the mother has a history of dislocated hip, the risk to the baby is increased to 1 per 25 nonbreech and 1 per 15 breech births.
- There is an increased association between developmental dysplasia of the hip (DDH) and foot deformities. The risk of DDH was highest for talipes calcaneovalgus (6.1%) and club foot (3.5%) in a recent study in Norway.
- Clinical exam maneuvers that assess hip dysplasia in children include:
  - Galeazzi test
  - Barlow test
  - Ortolani test
- Positive findings on the previously noted maneuvers should be confirmed by ultrasound (US) or x-rays and should not be repeated over and over.

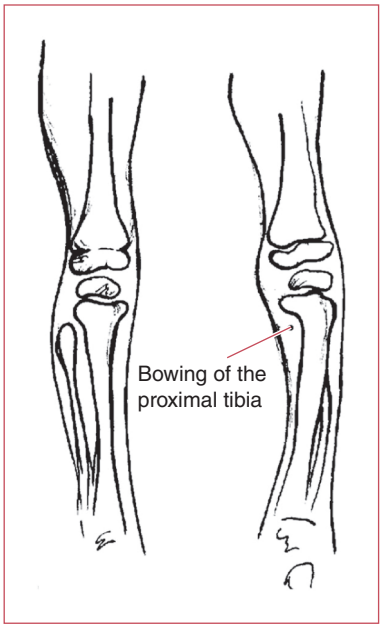


FIGURE 10–3 Blount's disease (tibia vara).

Galeazzi (Allis) Test (Figure 10–4)

- Flex hips and knees bilaterally, looking at the level of the knees. In the diagram, the level of the left knee is obviously lower, which usually indicates that hip dysplasia is present in this leg. Note: The same sign is seen in a congenital short femur, but this is a much less common finding.

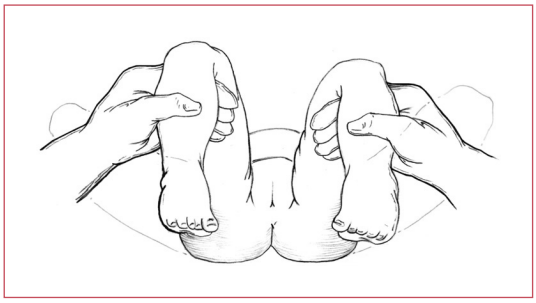


FIGURE 10–4 The Galeazzi (Allis) test.

Barlow and Ortolani Tests

- The Barlow and Ortolani tests are the classic maneuvers for congenital instability of the hip and are done in conjunction. These tests are now done under US observation to avoid missing bilateral hip dislocations.
- The *Barlow test* (**Figure 10–5**) is used to determine if a dislocated hip can be readily dislocated. At rest, the hip is reduced, and abduction is near normal or normal:
  - With the leg in a flexed and adducted position, push the femur posteriorly with the thumb.
  - If the hip dislocates posteriorly, as shown in the diagram, the Barlow test is positive and dislocation is palpable. Dislocation is verified with the Ortolani test, which reduces the dislocation.
- The *Ortolani test* (**Figure 10–6** and **10–7**) is used to determine if a dislocated hip can be readily reduced. If the hip remains dislocated for several weeks, hip abduction may become limited on the affected side:

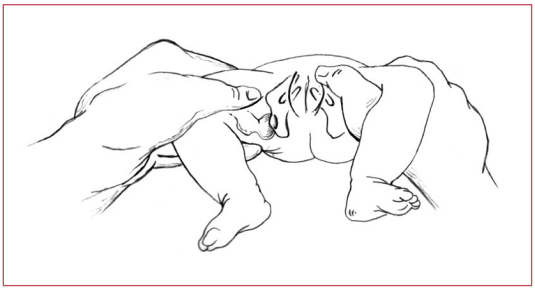


FIGURE 10–5 The Barlow test.

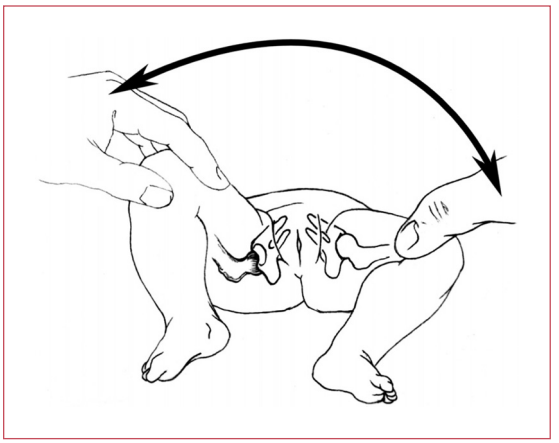


FIGURE 10–6 Ortolani test—Step 1: Femoral head is dislocated at rest; therefore, hip abduction is limited on the affected side.

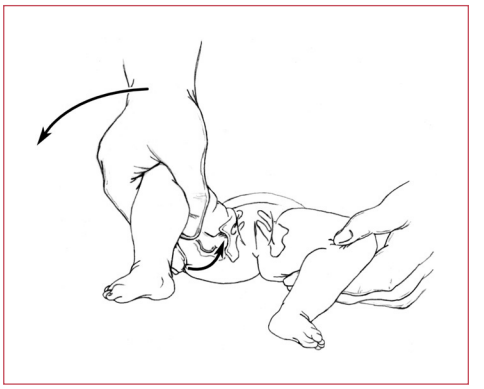


FIGURE 10–7 Ortolani test—Step 2: As abduction of the hip is attempted, reduction of the femoral head over the posterior lip of the acetabulum is performed by applying anterior pressure over the greater trochanter.

  - As the hip is gently abducted, the long finger over the greater trochanter pushes anteriorly to lift the femoral head over the posterior lip of the acetabulum to reduce the hip.
  - A positive Ortolani test is present when a palpable "clunk" is noted by the examiner as the hip reduces. A high-pitched "click" at full abduction is not a positive finding and is probably due to fascia lata slipping over the greater trochanter.

DIAGNOSIS AND TREATMENT

- If the diagnosis of hip instability is made in the first few months of life, closed reduction and use of a Pavlik harness or hip spica cast (to maintain hip reduction in 90- to 120-degree flexion and to limit hip adduction) for 3 to 4 months usually produces good results.
  - Care must be taken to avoid forced hip abduction in the brace or splint, as this may cause avascular necrosis (AVN) of the hip.
- If diagnosis is not made until walking age, surgery may be needed.

THE NECK

🕮 Congenital Torticollis ("Wry Neck")

- Congenital muscular torticollis affects 3.9% to 16% of infants, with the majority involving the right side. If treatment is started before 1 month of age, 98% of infants will achieve normal cervical range of motion (ROM) within 1.5 months.
  - The "olive sign" represents a soft, nontender enlargement of the sternocleidomastoid noted on physical examination. It is seen within the first 6 weeks and subsides within 4 to 6 months of age.
  - On exam, secondary deformities may include flattening of the ipsilateral face, contralateral occipital flattening, orbital asymmetry (plagiocephaly), and ipsilateral hip dysplasia.
- Torticollis can be the physical sign of an underlying problem usually due to muscular fibrosis, the presence of a cervical hemivertebra, or atlantoaxial rotary subluxation, which must be excluded. The head is tilted laterally toward one shoulder, with the chin rotated away from that shoulder:
  - In a child with *right* torticollis, the head is tilted to the *right shoulder* with the chin rotated to the left.
  - In a child with *left* torticollis, the head is tilted to the *left shoulder* with the chin rotated to the right.
- 🕮 The most common cause of congenital torticollis is fibrosis of the sternocleidomastoid muscle (SCM).

  - Suggested causes of this fusiform muscular swelling and fibrosis include birth trauma and ischemia due to the intrauterine position of the head and neck. The SCM is enlarged on the side toward which the head is laterally tilted.
  - Since there is a slightly higher incidence of DDH in children with muscular torticollis, the hips also need to be evaluated.
- The presence of a cervical hemivertebra is a less common cause of torticollis in the infant. Stretching exercises in this case would be of no benefit. A cervical hemivertebra can also lead to congenital cervical scoliosis, and surgical resection or fusion is performed if the cervical scoliosis increases with growth.

DIAGNOSIS AND TREATMENT

- X-rays of the cervical spine in congenital muscular torticollis will reveal rotation of C1–C2 related to the torticollis position or the presence of a cervical hemivertebra.
- 🕮 Conservative treatment should include the following:
  1. Mainstay of treatment is to stretch the contracted SCM 15 to 29 times per session, four to six times a day (at every diaper change).
     - With a *right torticollis*, tilt the child's head to the left (left ear toward left shoulder) and rotate the face to the right (chin to right shoulder).
     - With a *left torticollis*, tilt the child's head to the right (right ear toward right shoulder) and rotate the face to the left (chin to left shoulder).
  2. Position the crib so that the center of the room is on the side of the torticollis. This encourage the child's gaze toward the ipsilateral superior direction to stretch the ipsilateral neck muscles while strengthening the contralateral neck muscles.
  3. Put a mobile in the crib as follows:
     - For right torticollis, put the mobile to the right of the crib.
     - For left torticollis, put the mobile to the left of the crib.
     - If normal ROM is obtained by 1 year of age, facial asymmetry should resolve. Failure to regain full cervical ROM will lead to persistent facial asymmetry.
- Surgical intervention is considered when no improvement is shown by 18 to 24 months, with best results if performed when the child is <12 years old. Surgery includes resection of the fibrotic SCM:
  - In older children with persistent torticollis, surgical lengthening of the affected SCM is indicated.
  - Surgical fusion is performed to prevent increased scoliosis in patients with cervical hemivertebrae.

TRAUMATIC CONDITIONS

🕮 Subluxation of the Radial Head (Nursemaid's Elbow; Figure 10–8)

- The radial head and neck are displaced distal to the annular ligament.
- Results from longitudinal pull or sudden traction applied to the UE. Typical scenario is with a child <6 years old being lifted by the hand over a curb or object. There is sudden onset of pain, and the child subsequently refuses to move the arm. Hand function is normal, and elbow x-ray is usually read as normal.
- Reduction usually is achieved by supination and extension of the forearm.

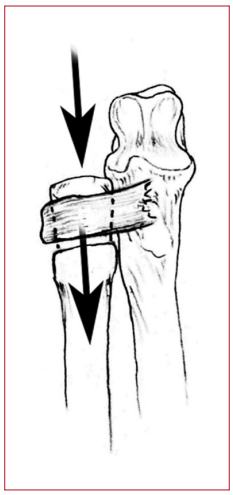


FIGURE 10–8 Subluxation of the radial head (nursemaid's elbow).

🕮 Medial Epicondylar Apophysitis (Little Leaguer's Elbow)

- Results from repetitive traction stress on the apophysis of the medial epicondylar ossification center of the humerus.
- Repetitive valgus stress on the elbow from activities such as throwing a baseball (especially pitching) commonly causes such injury.
- Treatment consists primarily of rest and instruction on proper body mechanics.

Osgood–Schlatter Disease (Figure 10–9)

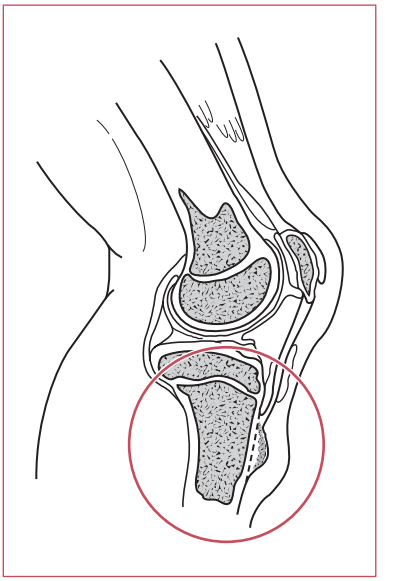


FIGURE 10–9 Osgood–Schlatter disease.

- Also known as Lannelongue's disease.
- 🕮 Osgood–Schlatter disease (OSD) is the most frequent type of knee pain in the adolescent population, affecting approximately 10% of adolescents. It is described as a traction apophysitis of the anterior tibial tubercle typically seen in active adolescent children, boys > girls (3:1). It typically affects boys 10 to 15 years old and girls 8 to 12 years old. OSD is a condition that results from chronic stress at the tibial tubercle from the patellar tendon attachment. It is a common, self-limited cause of anterior knee pain in adolescents, particularly those involved in sports activities that involve loading the knee (basketball, volleyball, and running). Up to 30% of patients have bilateral involvement.
- Pain results from inflammation/repeated microfractures in the apophyseal cartilage between the tibial tubercle and the secondary ossification center of the tibial tuberosity where the patella tendon attaches. These microfractures occur as the cartilage is less able to resist the stress from the patellar tendon from repeated knee extensions.
- Inflammation results, followed by pain, tenderness, and calcification in the area of cartilage involved.
- Physical exam reveals tenderness to palpation directly over the tibial tubercle and distal portion of the patellar tendon. The anterior tibial tubercle becomes prominent over time as a result of osseous healing of the tubercle.
- Diagnostic criteria consists of pain at the tibial tuberosity and pain on palpation over the tibial tuberosity. Imaging is the least likely diagnostic criteria, although it may assist in ruling out other diagnoses.
- Imaging:
  - The lateral x-ray of the knee may be normal or reveal soft tissue swelling anterior to the tibial tubercle, bony fragment over the anterior tibial tubercle, with loss of distinct infrapatellar fat pad pattern, and bony sclerosis.
  - US may be useful for confirmation of the diagnosis and also for follow-up. US can demonstrate soft tissue swelling, swelling of the cartilage, distal patellar tendon thickening with increased echogenicity (**Figure 10–10**), and fragmentation of the ossification center:

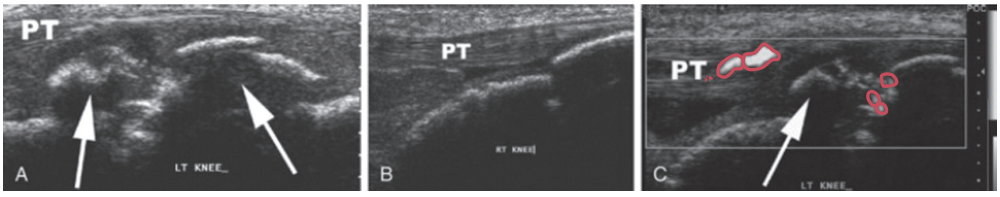


FIGURE 10–10 (A) Hyperechoic irregularity (arrows) associated with fragmentation of the tibial tuberosity apophysis common in OSD. (B) Normal contralateral knee for comparison. (C) Increased vascularity seen on Doppler imaging at insertion of affected patellar tendon.
OSD, Osgood–Schlatter disease; PT, patellar tendon.
Source: From Pope T, Bloem HL, Beltran J, et al. *Musculoskeletal Imaging*, 2nd ed. Elsevier, Saunders; 2014.

    - Hyperechoic irregularity (Figure 10–10A, arrows) associated with fragmentation of the tibial tuberosity apophysis common in OSD.
    - Normal contralateral knee for comparison (Figure 10–10B).
    - Increased vascularity seen on Doppler imaging at insertion of affected patellar tendon (Figure 10–10C).
  - MRI is a more sensitive and specific imaging modality and can demonstrate soft tissue swelling, patellar tendon thickening, avulsion/bony fragmentation of the ossification center, loss of the inferior angle of the infrapatellar fat pad, distended deep infrapatellar bursa (common finding), and edema of the anterior tibia bone.
- Treatment consists primarily of strenuous activity restriction (4–8 weeks), especially activities requiring deep knee bending or activities where the knee is in a flexed position (low bike seat). In addition, icing, oral anti-inflammatory medications, and cryotherapy can help with pain mitigation. PT should focus on strengthening the quadriceps, hamstrings, and gastrocnemius. Immobilization with a knee brace or cast has also been suggested. Surgery is reserved for pain that persists despite conservative therapy.
- OSD is a self-limited course with complete recovery with closure of the tibial growth place. Most adolescents are believed to be back to playing sports pain-free within 6 months. Research suggests that psychosocial factors (social exclusion, mental impact of being injured, and pain) seem to impact prognosis, although more research is required in this area.

🕮 NONTRAUMATIC HIP PAIN OR LIMP (TABLE 10–6)

- 🕮 **Transient (toxic) synovitis of the hip** is the most common cause of limping and pain in the hip of children. It is acute and nonspecific inflammation of the hip joint synovium.
- **Legg–Calvé–Perthes disease (LCPD)** (AVN of the proximal femur) is a result of rapid growth in relation to blood supply—the secondary ossification centers in the epiphysis are subject to AVN. More common in boys.
- **Slipped capital femoral epiphysis (SCFE; Figure 10–11)** is the most common hip disorder of preadolescent and adolescent children. It involves separation of the proximal femoral epiphysis through the growth plate (epiphysiolysis). More common in obese children, boys > girls.

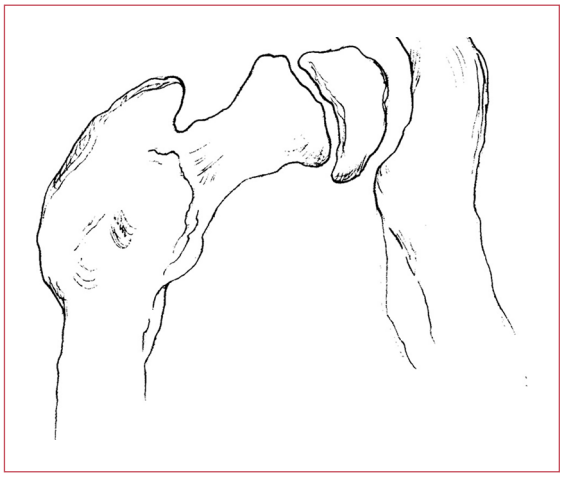


FIGURE 10–11 Slipped capital femoral epiphysis (SCFE).

SCOLIOSIS

- Scoliosis can be considered either functional or structural (Table 10–8).

Functional Scoliosis

- Occurs due to malpositioning or unilateral paraspinal muscle pull. This may be associated with back pain and muscle spasm.
- In this type of curve, there is no significant vertebral body rotation, and scoliosis is reversible.

TABLE 10–6 Causes of Nontraumatic Hip Pain or Limp

	ACUTE TRANSIENT/ TOXIC SYNOVITIS	LEGG–CALVÉ–PERTHES DISEASE	SLIPPED CAPITAL FEMORAL EPIPHYSIS
Etiology	Unclear	Avascular necrosis ossification in center of femoral head	Separation of proximal femoral epiphysis through the growth plate; obesity in 80% of children; family history of SCFE, endocrine/ metabolic disorder, delayed development of secondary sex characteristics
Incidence	2:1,000	0.4/100,000 to 29/100,000	10.8:100,000
Age at onset	3–6 years; boys > girls	4–10 years; boys > girls (4:1)	9–15 years; boys > girls; African American > Caucasian
Symptoms	Pain/limp Most common cause of acute hip pain in children	Pain/limp	Pain/limp; most common hip disorder in preadolescents–adolescents
Physical exam	Limited hip internal rotation	Pain in groin and radiates anterior/ medial thigh toward knee Decreased internal rotation, extension, and abduction	Decreased internal rotation, abduction, affected leg in external rotation Endomorphic habitus
WBC	Normal or slightly increased	Normal	Normal
ESR	Slightly increased	Normal	Normal
X-ray	Normal	Smaller ossified femoral head, sclerotic femoral head, widening of hip joint space	Physis (growth plate) on involved side wider and irregular; narrowing epiphysis (femoral head)
Treatment	Rest, NSAIDs, usually resolves in 3–5 days Full activity should be avoided until hip is pain-free	Conservative: Rest, abduction brace; the goal of treatment is to retain the normal spherical shape of the femoral head. Current therapy allows child to continue weight bearing, but with the femur in abducted position so that the head is well contained by the acetabulum. Surgical: Varus osteotomy	Treatment is surgical; pinning is preferred treatment, preventing further epiphyseal displacement by stabilizing the epiphysis with screws or pins
Prognosis	Good, <10% have second episode	Good prognosis in children detected earlier, i.e., <6 years old and with <50% of femoral head involvement; poor prognosis in children detected later, with involvement of the lateral portion of the femoral head (especially >40% of the structural part)	Variable

ESR, erythrocyte sedimentation rate; NSAID, nonsteroidal anti-inflammatory drug; SCFE, slipped capital femoral epiphysis; WBC, white blood cell.

Structural Scoliosis

- Not reversible.
- May be idiopathic, congenital, or acquired.
- **Idiopathic scoliosis (Table 10–7):**
  - Accounts for approximately 80% of patients with structural scoliosis.
  - Etiology is unclear.
  - Multifactorial condition with a genetic predisposition. There is an autosomal dominant component with incomplete penetrance.
  - Subdivided according to the age of onset with three well-defined periods:
    1. Infantile: 0 to 3 years old
    2. Juvenile: 4 years old to onset of puberty
    3. Adolescent: Puberty to just prior to closure of epiphyseal plates
  - Adolescent idiopathic scoliosis (AIS) has an annual incidence of 522.5 per 100,000 person-years. The incidence has decreased significantly after discontinuation of school screening in 2004.

TABLE 10–7 Clinical Features of Idiopathic Scoliosis

	INFANTILE	JUVENILE	ADOLESCENT
Incidence	Least common		Most common
Onset/diagnosis	<3 years old	4–10 years old	>11 years old
Ratio	Males > females	Males < females	Males < females; however, females worsen 8–10 times more frequently
Most common curve pattern	Left thoracolumbar	Right thoracic or double curve	Right thoracic 1; right thoracic/left lumbar 2

- **Congenital scoliosis:**
  - Results from abnormal spinal formation during embryonic development.
  - Most common presentation is with multiple hemivertebra or unilateral failure of segmentation in which one lateral half of the vertebra fails to form.
  - Congenital scoliosis is associated with a number of congenital anomalies:
    - About 30% of infants with congenital scoliosis have a congenital urinary abnormality, most commonly unilateral renal agenesis.
    - About one-third of children with congenital scoliosis also have a spinal cord abnormality.
    - In children with VACTERL (or VATER) syndrome (**V**ertebral defects, imperforate **A**nus, **C**ardiac defects, **T**racheo **E**sophageal fistula, **R**enal dysplasia, **L**imb abnormalities), congenital scoliosis is an additional finding.
  - All children with any congenital vertebral anomaly should have a renal US to assess for possible anomalies of the genitourinary (GU) system.
- **Acquired scoliosis—secondary to disease:**
  - Neuromuscular scoliosis:
    - Most commonly seen in cerebral palsy (CP), muscular dystrophy, spina bifida, and spinal muscular atrophy (SMA).
    - Scoliosis is uncommon in children with neuromuscular disease who are able to walk.
    - In CP, scoliosis is primarily seen in the spastic quadriplegic who is unable to stand or walk.
    - Scoliosis is unusual in Duchenne muscular dystrophy (DMD) until the child becomes a full-time wheelchair user.
    - Children with spina bifida who have no neurologic function below the thoracolumbar region have a high incidence of progressive scoliosis.
  - Connective tissue diseases

Scoliosis Classification (Table 10–8)

TABLE 10–8 Functional Versus Structural Scoliosis

FUNCTIONAL SCOLIOSIS	STRUCTURAL SCOLIOSIS
Muscle spasm Paraspinal strain Herniated disc (unilateral) Postural	**Congenital** Bar Block Hemivertebra or other body anomaly **Idiopathic** Adolescent (spinal cord or brainstem disease) Juvenile Associated with congenital heart disease **Acquired** Degenerative Posttraumatic (fracture) Overuse (repetitive microtrauma) Senile **Secondary (disease-related):** "Paralytic" neuromuscular disease (spinal muscular atrophy, muscular dystrophy, myelomeningocele, etc.), connective tissue disease (Ehlers–Danlos, chondrodysplasia, Marfan's, etc.)

Source: From O'Young B, Young MA, Stiens SA, eds. *PM&R Secrets*. Hanley & Belfus; 1997, with permission.

Evaluation of Scoliosis

ADAMS TEST (FORWARD BENDING TEST)

- The child bends forward with legs straight at the knees and hands together as if trying to touch the toes. The spine should be viewed from occiput down and sacrum up.
- Because scoliosis is a three-dimensional (3D) deformity, there is often rotation of the involved vertebrae as well as a lateral curvature (rotary scoliosis). This vertebral rotation can lead to a prominence of the posterior trunk that corresponds to the convex side of the curve.

COBB METHOD (FIGURE 10–12)

- The angle of a scoliotic curve is measured radiographically on a posterior-anterior (PA) or anterior-posterior view of the spine.
- One line is drawn along the superior endplate of the vertebra, tilted the most at the top of the curve, and a similar line is drawn along the inferior endplate of the vertebra, tilted the most at the bottom of the curve. Large scoliosis will allow these two lines to intersect, forming the angle "a." Otherwise, if the curve is not large enough, strike perpendiculars to form the angle "A."

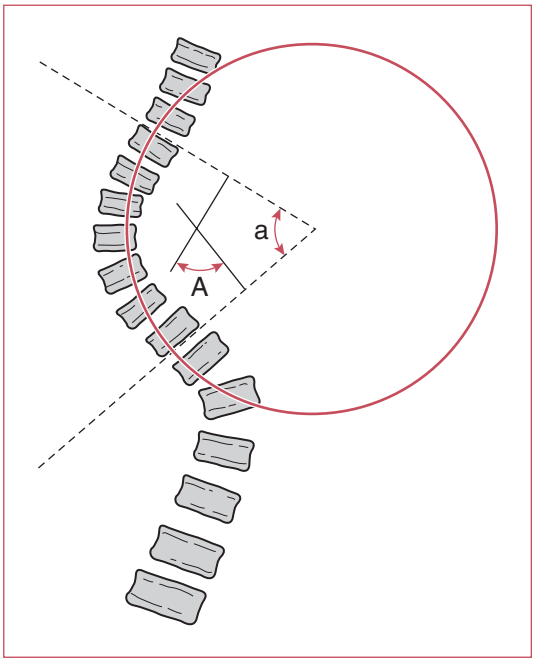


FIGURE 10–12 Cobb method.

PULMONARY FUNCTION TESTS

- If a thoracic scoliotic curve exceeds 50 to 60 degrees, pulmonary function may be compromised, resulting in abnormal pulmonary function tests (PFTs).
- Scoliosis typically results in a restrictive lung disease pattern with a decrease in total vital

capacity. The most common abnormality is a decrease in vital capacity related to a decrease in the size of the hemithorax on the convex side of scoliosis and loss of thoracic kyphosis. The higher the curvature, the more the lung is compressed on the convex side of the curvature. Spinal rotation results in lateral shifting of the ribs, which can further compress the lung (Tsiligiannis and Grivas, 2012).

Treatment of Scoliosis (Table 10–9)

- The goal of scoliosis treatment is early diagnosis, monitoring, and control of curvature progression during the growth spurt.
- During the period of rapid adolescent growth, progressive curves increase at the rate of approximately 1 degree per month.
- Studies have shown that if thoracic curvatures can be kept <50 degrees, and lumbar curvature is <40 degrees by the completion of growth, the likelihood of the curve increasing during adult life is small.

TABLE 10–9 Treatment of Scoliosis

DEGREE OF CURVATURE	IDIOPATHIC	NEUROMUSCULAR	CEREBRAL PALSY
1–20 degrees	Observation	Observation	Observation
20–40 degrees	Brace	Surgery (sooner if rapidly progressive)	
>40 degrees	Surgery		Surgery (may wait until 60 degrees or more in some cases)

Source: Modified from O'Young B, Young MA, Stiens SA, eds. *PM&R Secrets*. Hanley & Belfus; 1997, with permission.

SCHEUERMANN'S DISEASE (JUVENILE KYPHOSIS) (Tribus, 1998)

- Scheuermann's disease is a rigid kyphotic deformity in the thoracic > thoracolumbar spine and is reported to be the most common cause of back pain in children after spondylolysis with spondylolisthesis. It typically presents during adolescence.
- It occurs in 0.5% to 8% of the population, with an increased prevalence in males.
- Radiographically, Scheuermann's disease is diagnosed based on the Sorenson criteria of having three or more consecutive vertebrae with >5 degrees anterior wedging. Other radiographic findings can include irregular vertebral endplates, Schmorl's nodes (invagination of disc material into the spongiosum of the vertebral bodies), and increased intervertebral disc narrowing anteriorly.
- The cause is unknown, but theories include repetitive microtrauma and fatigue failure (osteochondrosis) of the immature vertebral bodies, genetic factors, and defective endplates.
- When Scheuermann's disease is associated with pain in the presence of one or more irregular vertebral bodies, physical exercises should be prohibited.
- Management depends largely on the degree of the kyphosis:
  - <50 degrees: Conservative, including PT, anti-inflammatories, behavior modification.
  - 50 to 75 degrees: Thoracic–lumbar–sacral orthosis (TLSO) brace may be required for pain control along with conservative measures of rest, ice, PT, and nonsteroidal anti-inflammatory drugs (NSAIDs).
  - Surgery recommended in cases with >75-degree curvature, refractory pain, or neurologic deficit due to severity of kyphosis.

SPONDYLOLISTHESIS

- Also read Chapter 4, Musculoskeletal Medicine, and Chapter 11, Pain Medicine, for additional information.
- Spondylolisthesis is a slippage of one vertebral body relative to the vertebral body below it.

- It is two to four times more common in males, but progression is more common in females.
- Patients typically present with back pain that can radiate to the buttocks or LEs. Symptoms typically worsen with lumbar extension and activities, such as standing and walking:
  - While rare, it can result in severe neurologic deficits such as cauda equina syndrome, particularly with an unstable or progressive spondylolisthesis. If the vertebra continues to slip, the spinal canal can become compromised, causing compression of the cauda equina.
- Imaging:
  - Radiographs (anterior-posterior, lateral, and oblique views) are usually the primary studies used to diagnose and manage spondylolisthesis in children and adolescents. Dynamic flexion–extension x-rays should be performed when there is suspicion of dynamic instability of the spondylolisthesis.
  - An MRI is recommended when neurologic deficits are noted.
- 🕮 In adults, degenerative spondylolisthesis is most common. However, in children, dysplastic and isthmic types are most frequent and occur most commonly at L5–S1 and then at L4–L5.
- 🕮 An **isthmic spondylolisthesis (Figure 10–13)** is the result of vertebral body slippage due to a spondylolysis (fracture of the pars interarticularis) and is the most common type of spondylolisthesis in children.
  - Most common type of spondylolisthesis.
  - Pars defect is at L5 in 71% to 95% of people, at L4 in 5% to 23%, and L1–L3 in 0% to 5%.
  - The frequency of spondylolysis in children is about 4.5%, in adolescents 6%, and increases to 12% in gymnasts.
- 🕮 A **dysplastic (congenital) spondylolisthesis (Figure 10–14)** involves a congenital malformation of the facet joints at the lumbosacral junction (congenital dysplasia of the sacrum or L5 neural arch) with pars elongation or attenuation (Wiltse et al., 1976). Spondylolysis may occur later as the slippage increases.
- Treatment of symptomatic spondylolisthesis:
  - If the slippage is <50%, treatment depends on the severity of symptoms. Anti-inflammatories and bracing are recommended in the initial acute stage. Transient restriction of activities followed by a course of PT with precautions to avoid spinal extension should then be performed.
  - Surgery is rarely needed with <25% slippage.
  - Generally, children with over 50% of slippage, progressive neurologic deficits, or an unstable spondylolisthesis require surgical fusion.

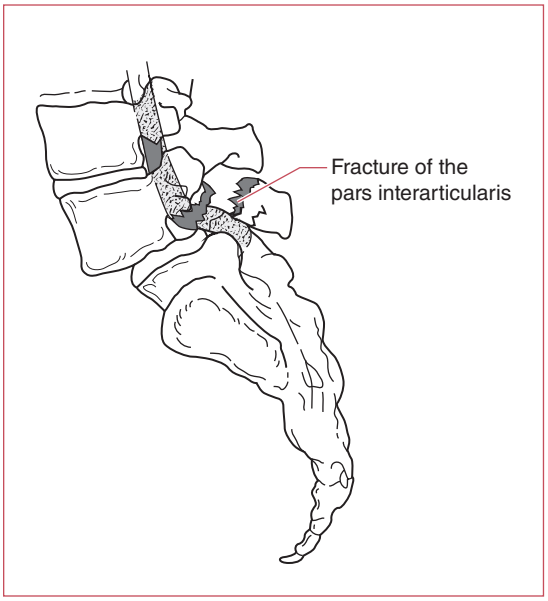


FIGURE 10–13 Isthmic spondylolisthesis resulting from pars fractures.

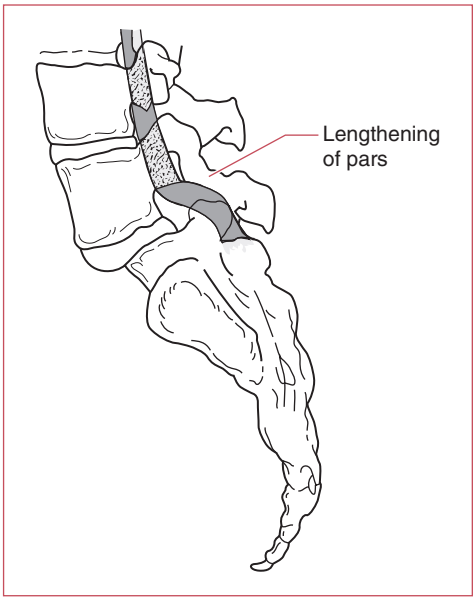


FIGURE 10–14 Dysplastic spondylolisthesis.

CONNECTIVE TISSUE AND JOINT DISEASE

JUVENILE IDIOPATHIC ARTHRITIS (TABLE 10–10)

- Also read Chapter 3, Rheumatology, for additional information.
- Formerly known as juvenile rheumatoid arthritis (JRA).
- 🕮 The most common connective tissue disease in children.
- It is the presence of arthritis lasting ≥6 weeks with onset <16 years of age.
- Childhood onset accounts for 5% of rheumatoid arthritis (RA) cases.
- The incidence is 2 to 20 cases per 100,000, with a prevalence of 16 to 50 cases per 100,000.
- The cause remains unknown; genetic predisposition, immunologic abnormalities, infection, and trauma are possible contributing factors.

Clinical Presentation

- There are seven main types of juvenile idiopathic arthritis (JIA):
  - **Polyarticular JIA:**
    1. Polyarticular rheumatoid factor (RF)-negative
    2. Polyarticular RF-positive
  - **Oligoarticular JIA**
  - **Systemic JIA**
  - **Enthesitis-related JIA**
  - **Psoriatic JIA**
  - **Undifferentiated JIA**

Polyarticular Juvenile Idiopathic Arthritis

- The polyarticular subtypes affect females > males.
- ≥5 joints involved in the first 6 months.

TABLE 10–10 Seven Types of Juvenile Idiopathic Arthritis

	POLYARTICULAR		OLIGOARTICULAR	SYSTEMIC	ENTHESITIS-RELATED	PSORIATIC	UNDIFFER-ENTIATED
	RF–	RF+					
% JIA patients	11%–28%	2%–7%	50%–60%	10%–20%	15%–20%	2%–15%	1%–7%
Sex	Female > male		Female > male	Both equally	Male > female	Both equally	Both equally
Joints	≥5 joints in the first 6 months		2–4 joints in the first 6 months; persistent and extended subtypes	≥1 joint	Arthritis + sacroiliitis, 🕮 HLA-B27+, boys >6 years old, or family history	Arthritis + psoriatic rash	Meets none or many of criteria from other subtypes
ANA	+20%–40%	-	+70%–80%	-	-	-	+ or -
Other key findings	5%–10% develop chronic uveitis	Severe arthritis >50%	Children <6 years old Ocular damage common 🕮 50% develop chronic iridocyclitis	🕮 10%–20% acute iridocyclitis	May progress to spondyloarthropathies Uveitis common	-	-

ANA, antinuclear antibody; HLA, human leukocyte antigen; JIA, juvenile idiopathic arthritis; RF, rheumatoid factor.

- There are two subgroups: RF-negative and RF-positive.
- **Polyarticular RF-negative:**
  - Comprises 11% to 28% of all patients with JIA.
  - 20% to 40% of cases are antinuclear antibody (ANA) positive; 5% to 20% are affected by chronic uveitis.
  - Children may complain of stiffness, but many do not complain of pain with inflamed joints.
  - Hip involvement occurs in 50% of children and is a cause of late disability from the erosion of the femoral head.
- **Polyarticular RF-positive:**
  - Occurs in 2% to 7% of children with JIA.
  - Findings include symmetric joint involvement, subcutaneous nodules, and erosive disease. Severe deforming arthritis is generally present within 5 years of onset.
  - This is the same as adult RF-positive disease.

Oligoarticular Juvenile Idiopathic Arthritis

- Accounts for the largest percentage of children (50%–60%) with JIA. Affects females > males.
- Involvement is limited to two to four joints in the first 6 months.
- There are two subgroups:
  - **Persistent**: Affects ≤4 joints throughout disease course.
  - **Extended**: Affects >4 joints after the first 6 months of disease.
- Presents in children <6 years old with asymmetric arthritis (usually lower limbs affected).
- Positive ANA titers in 70% to 80%. ANA+ indicates risk for iridocyclitis.
- Asymptomatic uveitis often develops within 4 years of diagnosis. As a result, these children require routine eye examinations.

Systemic Juvenile Idiopathic Arthritis

- 10% to 20% of JIA cases
- Characterized by arthritis in ≥1 joint, persistent fever (≥2 weeks duration), and ≥ 1 of the following:
  - Evanescent salmon-colored rash, hepatosplenomegaly, lymphadenopathy, serositis (pleural or pericardial).
- 5% to 8% of children with systemic JIA develop macrophage activation syndrome, a life-threatening complication presenting with persistent fever, lymphadenopathy, splenomegaly, and abnormal labs including a large decrease in ≥1 blood cell line, elevated liver function tests (LFTs), and abnormalities in clotting.

Enthesitis-Related Juvenile Idiopathic Arthritis

- Accounts for 15% to 20% of JIA cases, affecting males > females. Most common age at onset is 10 years old.
- Arthritis or enthesitis plus ≥2 of the following:
  - Sacroiliitis or inflammatory back pain, human leukocyte antigen (HLA-B27+), boys ≥6 years old, family history of spondyloarthritis, enteropathic arthropathy, reactive arthritis, or uveitis.
- Most common sites of enthesitis: Calcaneal insertion of Achilles tendon, plantar fascia, and tarsal area. Affects joints of the LE preferentially.
- Uveitis is common and occurs suddenly; it is symptomatic and unilateral.
- May progress to fulfill criteria for ankylosing spondylitis (AS), reactive arthritis, or arthritis associated with inflammatory bowel disease.

Psoriatic Juvenile Idiopathic Arthritis

- Accounts for 5% to 10% of JIA cases
- Arthritis and psoriatic rash OR arthritis plus ≥2 of the following: Dactylitis, nail pitting or onycholysis, psoriasis in a first-degree relative

Undifferentiated Juvenile Idiopathic Arthritis

- Accounts for 1% to 7% of JIA cases.
- Catch-all for those who do not meet the criteria for the previously mentioned subtypes or satisfy the criteria for more than one subtype.

Adult Rheumatoid Arthritis Versus Juvenile Idiopathic Arthritis

- Systemic features are more common in children.
- Adults have joint destruction earlier, whereas children have synovitis with erosive disease later.
- Children have large joints involved more frequently than adults with RA.
- Wrist and hand deviations differ from adults with RA; children have ulnar deviation at the wrist with loss of extension. Radial deviation of the fingers occurs at the metacarpophalangeal (MCP) joints with finger flexion.
- Tenosynovitis is more common in children than bursitis.
- Rheumatoid nodules occur less frequently in children than in adults.
- The cervical spine is more commonly involved in children than in adults.
- Arthritis in children tends to be ANA-positive, RF-negative, whereas adult RA is generally RF-positive.
- In children, periarticular bone demineralization is seen radiographically once 50% demineralization occurs.

Management of Juvenile Idiopathic Arthritis (Figure 10–15; Tables 10–11 and 10–12)

- The goal of management of JIA is to minimize deformity while maintaining as close to a normal lifestyle as possible.
- Treatment is aimed at inducing remission while minimizing medication toxicity.

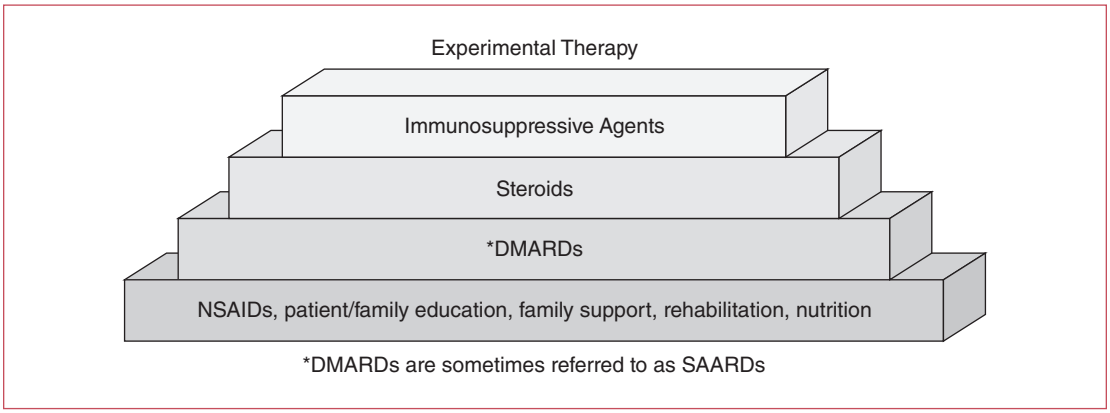


FIGURE 10–15 Pyramid approach to management of the child with JIA.
DMARDs, disease-modifying antirheumatic drugs; JIA, juvenile idiopathic arthritis; NSAIDs, nonsteroidal anti-inflammatory drugs; SAARDs, slow-acting antirheumatic drugs.

TABLE 10–11 Rehabilitation of the Child With Juvenile Idiopathic Arthritis

- Rest
- Splinting
- Passive ROM
- Active exercises for strengthening
- Adaptive equipment
- Functional training for ADLs and ambulation
- Postsurgical rehabilitation
- Nutrition
- Counseling: Family and child

ADLs, activities of daily living; ROM, range of motion.

TABLE 10–12 Drug Therapy in Juvenile Idiopathic Arthritis

DRUG	SIDE EFFECTS
Aspirin	Drowsiness, tinnitus, hyperventilation, concern of Reye's syndrome if used during varicella or influenza, reduced platelet function, GI irritation
Naproxen	GI irritation, cutaneous pseudoporphyria, cutanea tarda
Ibuprofen	GI irritation, rash, aseptic meningitis
Tolmetin	GI irritation
Indomethacin[a]	Headache, epigastric pain, difficulty paying attention
Diclofenac[a]	Mild GI effects
Piroxicam[a]	Side effects not established in pediatric population
Gold salts	Mucosal ulcers, rash, proteinuria, nephropathy, leukopenia, thrombocytopenia, anemia
Auranofin[a]	GI irritation, rash
Hydroxychloroquine	Macular degeneration
d-Penicillamine	Bone marrow suppression, renal, rash, autoimmune, proteinuria
Sulfasalazine[a]	GI irritation, rash, hypersensitivity, renal toxicity, headache
Methotrexate[a]	Avoid use with NSAIDs because it may potentiate bone marrow suppression, GI side effects, hepatotoxicity
Azathioprine[a]	GI irritation, side effects liver, side effects dose-related leukopenia
Cyclophosphamide[a]	Alopecia, nausea, vomiting, bladder side effects, pulmonary fibrosis, leukopenia, thrombocytopenia
Cyclosporine	Immunosuppression, hypertension, renal insufficiency
Corticosteroids	Growth failure, adrenal suppression, osteopenia, cushingoid appearance, avascular necrosis, weight gain, cataracts, psychosis, myopathy
Prednisone	Same side effects as for corticosteroids
Pulse steroid methylprednisolone	Same side effects as for corticosteroids

[a]Not approved for use in children.
GI, gastrointestinal; NSAIDs, nonsteroidal anti-inflammatory drugs.
Source: From Molnar GE, Alexander MA. *Pediatric Rehabilitation*. 3rd ed. Hanley & Belfus; 1999, with permission.

- The American College of Rheumatology regularly updates their recommendations regarding clinical practice guidelines. The major changes to the 2021 JIA guideline are the use of bDMARDs (biological disease-modifying antirheumatic drugs) as initial treatment or following inadequate response to a short course of NSAIDs. The addition of csDMARDs (conventional synthetic disease-modifying antirheumatic drugs) is recommended for persistent synovitis despite treatment with bDMARDs. Triamcinolone hexacetonide was recommended for oligoarthritis. The panel supported the use of oral glucocorticoids and NSAIDs, sparingly and as a bridge until more definitive treatment is available. In addition, the guidelines support the use of PT and occupational therapy (OT) services, a well-balanced healthy diet, laboratory monitoring while on antirheumatic medications, the importance of immunizations, and shared decision-making among patients and their caregivers. Unfortunately, the recommendations are not supported by high-quality evidence, furthering the need for more high-quality evidence to support or refute the current management of JIA.

MEDICATIONS (TABLE 10–12)

- **Aspirin and NSAIDs:**
  - Aspirin is used less frequently because of the risk of Reye's syndrome with influenza and Varicella infections.
  - Of the NSAIDs, naproxen, ibuprofen, and tolmetin are approved for use in children. More than 50% of children improve with NSAID therapy. Treatment is continued for 1 to 2 years after suppression of disease activity.
- **Disease-modifying antirheumatic drugs (DMARDs):**
  - Second line of drug therapy that includes methotrexate, gold salts, antimalarials, D-penicillamine, leflunomide, tocilizumab (interleukin [IL]-6 inhibitor), abatacept (T-cell inhibitor), anakinra (calcineurin inhibitor), and sulfasalazine. However, studies have not demonstrated that disease progression is modified with DMARDs.
  - 60% to 70% of children receiving gold salts showed improvement. Side effects include skin rash, proteinuria, and bone marrow suppression. Oral gold (auranofin) has been found to be safe for use in children but less efficacious than intramuscular (IM) administration.
  - Hydroxychloroquine is the most commonly used antimalarial drug for JIA.
  - Oral D-penicillamine induces remission in 60% to 70% of cases.
- **Cyclosporine:**
  - Blocks production of IL-2 and the proliferation of synovial T-cells.
  - May have disease-modifying as well as anti-inflammatory effects.
- **Azathioprine:**
  - Effective treatment for children with refractory disease and can reduce the use of steroids
  - It was not found to alter the cause of iridocyclitis.
- **Corticosteroids:**
  - Systemic corticosteroids reduce symptoms but do not cause remission. Used sparingly to control inflammation in order to avoid long-term side effects.
  - Intra-articular steroids can suppress synovitis.

Additional Aspects of Managing Juvenile Idiopathic Arthritis

- Joint pain is caused by stretching of the capsule. Pain can be assessed using the child visual analogue scale (VAS) or the Varni/Thompson Pediatric Pain Questionnaire, which takes into account the child's cognitive level.
- Heat can reduce stiffness, increase tissue elasticity, and decrease pain and muscle spasms. Aquatherapy can also be used to manage JIA. Water temperature should be 90°F to 100°F. There remains concern that heat can increase inflammation and accelerate the disease process that leads to articular destruction. Superficial heat (e.g., moist heat, hot packs) can provide heat to tissues up to 1 cm deep. US can provide deep heat; however, it has the potential to affect the growth plates in children and should therefore be used with caution in this population. Cold can provide pain relief, increase the pain threshold, and decrease muscle spasm and swelling by vasoconstriction.
- Splinting: The UE is splinted in a functional position with the wrist in 15- to 20-degree extension, the fingers flexed (25% at the MCP joint and a few degrees at the proximal interphalangeal [PIP] joints), with the thumb in opposition.
- In an inflammatory joint, traditionally passive ROM activities should not be performed.

Specific Joints of Involvement in Juvenile Idiopathic Arthritis

- Cervical spine—involved more often in children than in adults. Subluxation of the atlantoaxial joint can occur when its involvement leads to erosion of the transverse ligament.
- Temporomandibular joint (TMJ)—involved in up to 50% of children with JIA. Micrognathia results from reduced mandibular growth.
- Shoulder—uncommon, occurring in only 8% of children with JIA. Children lose abduction and internal rotation, in contrast to adults who lose external rotation.

- Elbow—≥90% of elbow flexion is needed for activities of daily living (ADLs).
- Wrist—common in children. Early loss of wrist extension leads to progression of a wrist flexion contracture. (Note: In children, the wrist deviates in the ulnar direction.)
- Hand—swan-neck deformity: Hyperextension at the PIP joint with MCP and distal interphalangeal (DIP) flexion is more common in adults. Boutonnière deformity: Flexion at PIP joint with hyperextension at the MCP and DIP. (Note: Radial deviation of the fingers occurs at the MCP joint.)
- Hip—occurs in 50% of children with polyarticular arthritis. Hips develop flexion contractures with internal rotation and adduction compared with adults who commonly exhibit hip external rotation and abduction.
- Knee—holding the knee in 30 degrees of flexion minimizes intra-articular pressure.
- Ankle/Foot—involvement at the metatarsophalangeal joint causes pain and decreased push-off, resulting in flat foot gait.

Functional Outcome in Rheumatoid Arthritis (Table 10–13)

- 31% of children have severe limitations in Class III or IV. Remission occurs in up to two-thirds of children, whereas adults usually progress.
- Poor outcome is related to:
  - Delay in treatment
  - Later age at disease onset
  - Longer duration of disease as remission is unlikely after >7 years in time
  - RF-positive status
  - Unremitting course
  - Multiple small joint involvement
  - Early appearance of erosion
  - Hip involvement
- Death occurs in 2% to 4% of children.

JUVENILE-ONSET SERONEGATIVE SPONDYLOARTHROPATHIES

- Juvenile-onset seronegative spondyloarthropathies include HLA-B27-associated syndromes in children <16 years old with findings related to arthritis, enthesitis, and tenosynovitis involving joints of the LE and spine, and sacroiliac (SI) joint.
- RF and ANA are negative (RF-negative = seronegative).
- They are usually more common in boys than girls.
- Treatment with tumor necrosis factor inhibitor (TNFi) resulted in improved active joint and active enthesitis counts, and patient-reported pain 6 months post initiation. However, patients were still found to have active disease 1 year following initiation with TNFi.
- Includes the following three entities:
  - Ankylosing spondylitis
  - Reactive arthritis (formerly Reiter's syndrome)
  - Enteropathic arthritis (arthritis with irritable bowel disease [IBD])

TABLE 10–13 American College of Rheumatology Revised Criteria for Classification of Functional Status in Rheumatoid Arthritis

Class I	Completely able to perform usual activities of daily living (self-care, vocational, avocational).
Class II	Able to perform usual self-care and vocational activities but limited in avocational.
Class III	Able to perform usual self-care activities but limited in vocational and avocational.
Class IV	Limited in ability to perform usual self-care, vocational, and avocational activities.

Source: From Hochberg MC, Chang RW, Dwosh I, et al. The American College of Rheumatology 1991 revised criteria for the classification of global functional status in rheumatoid arthritis. *Arthritis Rheum*. 1992;35:498–502. doi:10.1002/art.1780350502, with permission.

1. Ankylosing Spondylitis

- Incidence: 2 per 100,000 in the United States and occurs more often in boys older than 8 years old; 90% of Caucasian patients with AS are HLA-B27-positive.
- Prevalence ranges between 0.2% and 0.5%.
- Cause: Unknown. There is a strong genetic susceptibility. Axial spinal symptoms (spine and SI joint) and radiographic sacroiliitis develop.
- Children often have peripheral joint involvement (82%), with LE and hip most often involved.
- Enthesitis, pain at the insertion of tendon to bone, occurs more commonly in children than adults.
- Up to 27% have associated uveitis.
- Radiographic findings of bilateral SI joint involvement are necessary for the definitive diagnosis.
- Hip disease is an indicator for poor outcome.

2. Reactive Arthritis (Formerly Reiter's Syndrome)

- Characterized by a triad of asymmetric arthritis, conjunctivitis, and urethritis/gastroenteritis (inflammation of joint(s), eyes, and GU/gastrointestinal [GI]).
- Oligoarthritis of the knee or ankle is most common.
- Overall uncommon in children. More frequent in boys >8 years old.
- Postinfectious or reactive cause secondary to GI or GU infection typically with *Chlamydia trachomatis, Chlamydia pneumoniae, Salmonella, Shigella flexneri*, or *Yersinia enterocolitica*.

3. Enteropathic Arthritis

- Inflammatory arthritis associated with GI pathology, commonly with IBD.
- Occurs in 10% to 20% of children with ulcerative colitis and Crohn's disease.
- No gender predilection.

SYSTEMIC LUPUS ERYTHEMATOSUS (TABLE 10–14)

- Chronic systemic autoimmune disease occurring with episodic inflammation and vasculitis associated with a positive ANA. Cause is unclear.
- Incidence is 5.1 per 100,000, with 20% of cases beginning in childhood
- Females are predominantly affected 8.7:1.2 in all age groups. A closer ratio exists in prepubertal patients.
- Diagnosis is likely if at least ≥4 of 11 diagnostic criteria for systemic lupus erythematosus (SLE) are present at any time.
  - The presence of four criteria has 90% sensitivity and 98% specificity.
- One-third of children have an erythematous rash over the bridge of the nose and cheeks in a butterfly distribution (malar rash).

TABLE 10–14 11 Diagnostic Criteria for Systemic Lupus Erythematosus (SLE)[a]

1. Malar rash
2. Discoid lupus rash
3. Photosensitivity
4. Oral or nasal mucocutaneous ulceration
5. Nonerosive arthritis
6. Nephritis
7. Encephalopathy
8. Pleuritis or pericarditis
9. Cytopenia
10. Positive immunoserology: LE cells, anti-ds-DNA antibodies, anti-Sm antibodies, false+ test for syphilis
11. Positive ANA titer

[a]Four or more positive criteria required for clinical diagnosis.
ANA, antinuclear antibody; anti-ds-DNA, anti-double-stranded DNA; LE cells, lupus erythematosus cells.
Source: From Molnar GE, Alexander MA. *Pediatric Rehabilitation*. 3rd ed. Hanley & Belfus; 1999, with permission.

- Nonerosive arthritis and joint deformities without bony erosions may develop with chronic SLE.
- Nephritis is present in 75% and is the main factor in determining outcome in children.
- 10-year survival is 90%.
- Hematuria, proteinuria, persistent hypertension, pulmonary hypertension, chronic active disease, and biopsy-proven diffuse proliferative glomerulonephritis are associated with a poor outcome.

JUVENILE DERMATOMYOSITIS

- Multisystem inflammatory disease of unknown cause, primarily involving the muscle, skin, and subcutaneous tissues.
- 🕮 Clinical features include histologic presence of vasculitis, the onset of calcinosis, and lack of association with childhood malignancy.
- It occurs more commonly in girls between 5 and 14 years old.
- Higher incidence in juvenile dermatomyositis of systemic vasculitis and calcification in soft tissues (muscle, subcutaneous skin), and lipodystrophy.
- Diagnostic features of juvenile dermatomyositis include:
  - Proximal muscle weakness.
  - Characteristic periorbital heliotropic rash.
  - Elevated muscle enzymes.
  - Electromyography (EMG) evidence of inflammatory myopathy.
  - Evidence of vasculitis or chronic inflammation on muscle biopsy.
  - Other signs/symptoms may include fever, muscle tenderness and pain, malaise and weight loss, arthralgia and arthritis, dyspnea, dysphagia, myocarditis with abnormal EKG, and +ANA.
- The clinical course of this disease is variable.

Treatment Options

- Corticosteroid therapy is indicated in acute or active disease. Prednisone therapy with a slow taper over 2 years once muscle enzyme elevations have normalized is standard.
- Immunosuppressive agents may be used in refractory cases.
- PT is essential to prevent contractures and is instituted once the muscles are less inflamed.
- The prognosis is good with <7% mortality.
- Those with the chronic form of disease have calcinosis and functional disability in adulthood.

SCLERODERMA

- A chronic, systemic connective tissue disease that chiefly affects the skin, internal organs, and walls of blood vessels. The cause is unclear.
- Although uncommon in children, it affects girls > boys with fibrosis of involved tissue.
- Rare in children. Average age of onset in children is between 8 and 10 years old with duration of 7 to 9 years.
- Treatment with methotrexate in combination with steroids has led to cessation of disease progression, improvement in inflammatory skin changes, partial reversal of sclerosis and dermal atrophy, as well as improvements in alopecia. The most significant improvement was noted in the first 3 months of treatment. No added benefit was seen with thermography.

Types of Scleroderma

1. MORPHEA

Localized form of scleroderma that is limited to skin involvement, small lesions occur with minimal sclerosis (guttate morphea), self-limiting over 2 to 3 years.

2. SYSTEMIC SCLEROSIS

Characterized by Raynaud's phenomenon; symmetric cutaneous involvement; loss of joint function, involvement of the lungs, GI tract, and kidneys, and pulmonary and renal complications. GI, pulmonary and renal complications are causes of death in these children.

3. OVERLAP SYNDROMES

Includes mixed connective tissue diseases (MCTD), which have features of SLE, RA, dermatomyositis, and scleroderma.

4. OTHER

CREST Syndrome (**C**alcinosis, **R**aynaud's, **E**sophageal dysfunction, **S**clerodactyly, **T**elangiectasia)

INFECTIOUS ARTHRITIS

Lyme Disease

- Cause: *Borrelia burgdorferi*, a spirochete transmitted by the deer tick, *Ixodes dammini.*
- Incidence: 7.21 per 100,000.
- Initial phase is characterized by fever, fatigue, headache, arthralgias, myalgia, and stiff neck.
- Erythema migrans is the characteristic round, red skin lesion with central clearing.
- Late phase: Characterized by arthritis, cardiac disease, and neurologic disease.
- Cardiac manifestations of heart block occur in 5% to 10% of children and chronic neurologic manifestations occurs in 15%.
- Bell's palsy is seen more frequently in children than in adults.
- In 85% of children, the arthritis resolves before the end of the initial treatment, but a chronic inflammatory phase develops in 10%.
- Treatment is with antibiotic therapy: Doxycycline, Amoxicillin, Erythromycin (late disease IV—Ceftriaxone).
- Researchers are currently evaluating options for passive or active immunization against Lyme. A Lyme vaccine (VLA15) has received fast-track designation by the Food and Drug Administration (FDA) and is being evaluated in a phase 2 clinical trial.

Rheumatic Fever

- Rheumatic fever occurs in children >4 years old, with boys and girls affected equally.
- Arthritis presents with pain, swelling, warmth, and decreased joint ROM in large joints, more commonly the knees, elbows, ankles, and wrists.
- Associated findings include carditis, fever, rash, chorea, and nodules.
- There often is a history of a prior streptococcal infection. Diagnosis is clinical by the Jones criteria (Table 10–15).

TREATMENT

- Management includes anti-inflammatory medications (salicylates, corticosteroids) and PT.

TABLE 10–15 Jones Criteria: Diagnosis of Rheumatic Fever

MAJOR	MINOR	PRECEDING GROUP A STREPTOCOCCAL INFECTION
Carditis	Fever	Throat culture
Polyarthritis	Arthralgia	Rapid streptococcal antigen
Chorea	Elevated ESR, or CRP	Elevated streptococcal antibody
Erythema marginatum	Prolonged PR interval	
Subcutaneous nodules		

Note: Two of the major criteria, or one major and two minor criteria, are required for diagnosis with evidence of preceding streptococcal infection.
CRP, C-reactive protein; ESR, erythrocyte sedimentation rate.
Source: From Molnar GE, Alexander MA. *Pediatric Rehabilitation*. 3rd ed. Hanley & Belfus; 1999, with permission.

PROGNOSIS

- Arthritis does not result in long-term morbidity, but prognosis is related to the extent of cardiac involvement.

Septic Arthritis (Table 10–16)

- Occurs most often in children <2 years old, with boys > girls, and occurs through hematogenous spread into the joint.
- Bacterial septic arthritis accounts for 6.5% of arthritis in children. Monoarticular involvement is most common.
- Common pathogens are *Haemophilus influenzae* and *Staphylococcus aureus*.

TABLE 10–16 Causes of Septic Arthritis in Children

AGE	MOST COMMON BACTERIA
Newborn	*Staphylococcus aureus* + *Group B Strep*
2 months to 2 years old	*Haemophilus influenzae*
More than 2 years old	*Staphylococcus aureus* + *Group B Strep*
Sexually active adolescents	Gonococcal disease

HEMOPHILIAS

Hemophilias are the most common and serious of the congenital coagulation disorders. They are associated with genetically determined deficiencies of Factor VIII, IX, or XI (gene is carried on the X chromosome).

Classification

- Hemophilia A (classic hemophilia): Factor VIII deficiency
- Hemophilia B (Christmas hemophilia): Factor IX deficiency
- Hemophilia C: Factor XI deficiency
- 🕮 The hallmark of hemophilia is **hemarthrosis** (hemorrhage into the joints causing pain, inflammation, swelling, and limited joint mobility).
  - This may be induced by minor trauma but can also occur spontaneously.
  - Repeated hemorrhages can result in degenerative changes with osteoporosis, muscle atrophy, and ultimately a fixed nonfunctional joint.
- Disease severity:
  - Mild disease presents with >5% factor activity.
  - Moderate disease presents with >1% factor activity.
  - Severe disease presents with <1% factor activity.

Treatment

- Prevention of trauma (avoid contact sports and high-impact activities).
- Avoid aspirin and other platelet-affecting drugs because they may provoke severe hemorrhage.
- During bleeding episodes:
  - Factor VIII replacement therapy via intravenous (IV) infusion is used to increase ability for hemostasis.
  - Factor VIII replacement therapy is essential to preventing pain, disability, or life-threatening hemorrhage.
  - Emicizumab, a bispecific antibody that mimics factor VIII, is being evaluated as prophylaxis in children with hemophilia. In a retrospective review at Texas Children's Hospital from 2018 to 2021, the mean annualized bleeding rate decreased from 3.61 to 0.44 in 28 patients treated with Emicizumab prophylaxis.

  - When a hemophiliac child has significant bleeding:
    - Factor VIII should be given as soon as possible.
    - Local measures should include application of cold and pressure.
    - Initially immobilize. Within 48 hours, passive exercises should begin to prevent joint stiffness and fibrosis.
    - Joint aspiration is controversial (tamponade effect is lost).
    - There is compelling evidence that early treatment with Factor VIII concentrates will reduce disability and deformity. Parents or older patients can be trained to give IV concentrates at home.
- Complications of treatment with Factor VIII replacement:
  - Abnormalities of hepatic enzyme activities are found in 50% of patients.
  - 30% of hemophiliacs are currently infected with HIV from factor transfusions.
  - Hepatitis B and C are also transmissible by factor concentrate.

KAWASAKI DISEASE (INFANTILE POLYARTERITIS)

- Systemic vasculitis that affects young children, with an annual incidence of 9 to 20 per 100,000 in children under 5 years old. The cause is unknown.
- 80% of affected children are <4 years old, boys > girls.

Diagnostic Criteria in Kawasaki Disease (Molnar and Alexander, 1999)

- High-grade fevers lasting >5 days
- Strawberry tongue
- Bright red, chapped lips
- Pharyngeal erythema
- Conjunctival injection
- Edema of the hands or feet
- Erythema of the palms or soles with desquamation in later stages
- Truncal rash
- Cervical lymphadenopathy

PEDIATRIC BURNS

EPIDEMIOLOGY

- Burn injuries account for about 180,000 deaths annually and are the fifth most common cause of nonfatal childhood injuries.
- The World Health Organization (WHO) launched a Global Burn Registry (GBR) to collect and analyze burns worldwide.
  1. 42% were children, with the mean age being 5.3 years old.
  2. Children aged 1 to 5 years old accounted for 62% of the pediatric cohort and 26% of all burn injuries.
  3. School-aged children, aged 6 to 12 years, accounted for 20%, followed by adolescents, aged 13 to 18, who accounted for 13%.
  4. Males were reported more than females: 60% vs. 40%.
  5. Scald injuries represented 62% of pediatric burns, with the highest incidence occurring in toddlers aged 1 to 5 (80%).
     - 🕮 **Scald burns** are the single most common cause of pediatric burn injury. Immersion scald burns have been associated with child abuse or neglect.
- Increased burn size raises the risk of mortality in children.
- Inhalation injury is an important predictor of burn mortality.

- 16% of all burn injuries are nonaccidental, and 50% of these are a result of abuse.
- Flame burns resulting from playing with matches, gasoline, firecrackers, and flammable aerosols continue to occur in the 6- to 14-year-old group. Education programs are geared toward this age group.
- Wall outlet-caused electrical injuries represent <15% of all pediatric electrical injuries.

BURN CLASSIFICATION

- Calculation of burn size is relative to total body surface area (TBSA) and varies depending on age. The modified Lund and Browder chart is most often used in estimating the extent of burn size in children (Table 10–17).

TABLE 10–17 Child Burn Size Estimation Chart (% Body Surface Area)

BURN AREA	AGE IN YEARS				
	Birth to 1	1–4	5–9	10–14	15
Head	19	17	13	11	9
Neck	2	2	2	2	2
Anterior trunk	13	13	13	13	13
Posterior trunk	13	13	13	13	13
Buttock	5	5	5	5	5
Genitalia	1	1	1	1	1
Upper extremity (each)	9.5	9.5	9.5	9.5	9.5
Lower extremity (each)	14	15	17	18	19

Source: Adapted from Alexander MA, Matthews DJ, eds. *Pediatric Rehabilitation*. 5th ed. Demos Medical Publishing; 2015, with permission.

Rule of 9s (Figure 10–16)

- The rule of 9s is another frequently used method of estimating burn size. It is modified for use with children.
- 9% is taken from the legs and added to the head of a child <1 year old. For each subsequent year, 1% is returned to the legs until 9 years old, at which time the head is in proportion to the adult.

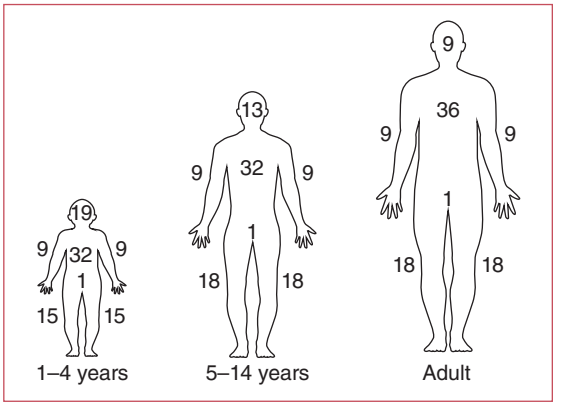


FIGURE 10–16 The rule of 9s.

Classification by Severity (Figure 10–17)

- The depth of thermal injury through epidermis and dermis determines classification.

SUPERFICIAL BURNS (FORMERLY FIRST DEGREE)

- Typified by sunburn; they are dry, warm, painful, and hypersensitive. They heal without treatment.

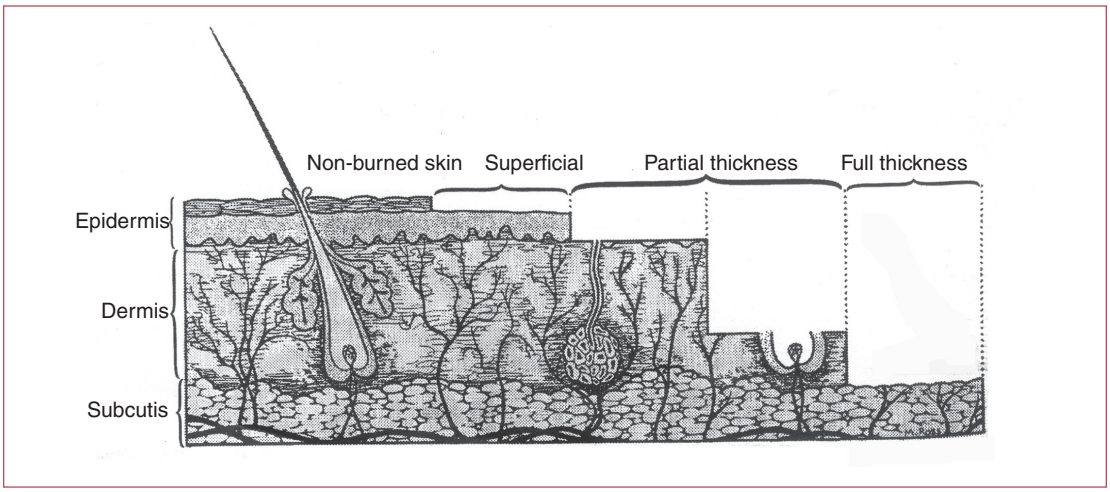


FIGURE 10–17 Classification of burns.
Source: From Alexander MA, Matthews DJ, eds. *Pediatric Rehabilitation*. 5th ed. Demos Medical Publishing; 2015, with permission.

PARTIAL THICKNESS BURNS (FORMERLY SECOND DEGREE)

- Burns affect the epidermis, and variable (but not complete) elements of the dermis. Partial thickness burns can be superficial, resulting in burns that are red, painful, and potentially blistered; or deep, resulting in dry, white, and hyposensitive burns. Treatment depends on depth of damage.

FULL THICKNESS BURNS (FORMERLY THIRD DEGREE)

- Affects all epidermal and dermal elements, and are anesthetic, white, avascular, dry, and leathery appearing. Skin grafting helps ameliorate the uneven and hard fibrotic scar formation as healing proceeds.
- The American Burn Association established a guideline for hospitalization of a burn patient based on the 5–10–20 rule (see next section for more detail).

INDICATIONS FOR HOSPITALIZATION (Molnar and Alexander, 1999)

- >5% full thickness injury
- >10% TBSA in children or elderly
- >20% TBSA
- Any burns to the eyes, ears, face, hands, feet, genitalia
- All inhalation injuries
- All electrical burns
- All burns complicated by other medical problems
- Any burns associated with concomitant trauma

POSITIONING IN THE PEDIATRIC BURN PATIENT (TABLE 10–18)

Proper Positioning

- Proper positioning of the burned child during the acute phase of care is essential in preventing contractures and deformity and controlling edema. Typically, the position of comfort is one that promotes deformity and should be avoided.
- Remember, in burn patients, COMFORT = CONTRACTURE!

TABLE 10–18 Proper Positioning of a Pediatric Burn Patient

AREA INVOLVED	CONTRACTURE PREDISPOSITION	CONTRACTURE PREVENTING POSITION
Anterior neck	Flexion	Extension, no pillows
Anterior axilla	Shoulder adduction	90-degree abduction, neutral rotation
Posterior axilla	Shoulder extension	Shoulder flexion
Elbow/forearm	Flexion/pronation	Elbows extended, forearm supinated
Wrists	Flexion	15- to 20-degree extension
Hands		
-MCP	Hyperextension	70- to 90-degree flexion
-IP	Flexion	Full extension
-Palmar burn	Finger flexion, thumb opposition	All joints full extension, thumb radially abducted
Chest	Lateral/anterior flexion	Straight, no lateral or anterior flexion
Hips	Flexion, adduction, external rotation	Extension, 10-degree abduction, neutral rotation
Knees	Flexion	Extension
Ankles	Plantar flexion	90-degree dorsiflexion

IP, interphalangeal; MCP, metacarpophalangeal.
Source: From Alexander MA, Matthews DJ, eds. *Pediatric Rehabilitation*. 5th ed. Demos Medical Publishing; 2015, with permission.

BURN REHABILITATION PRINCIPLES

Splinting

- Positioning splints with goals of contracture prevention are useful adjuncts for the pediatric burn victim.
- Splinting is generally initiated at the first indication of developing skin tightening.
- In contrast to adults and adolescents, small children do not tend to lose strength or joint mobility when immobilized in splints for extended periods, provided that the splints are removed for regular exercise or an activity session.

Range of Motion

- Once the child is medically stable, ROM should move from gentle, repetitive action to more aggressive stretching.
- Active or active assisted ROM is preferable to passive. However, it may be necessary to combine both.

Activities of Daily Living

- Children who have sustained burn injury should be encouraged to participate in ADLs as soon as they are medically stable.

Exercise

- Children frequently experience loss of strength and motor function as a result of their initial burn injury.

- Exercise programs should emphasize flexibility, strength, and endurance.
- Ambulation should begin as soon as the child's physical condition allows, often by 48 to 72 hours after injury, when vital signs are stable and fluid resuscitation is complete.

Physical Modalities

- Fluidotherapy and paraffin may be helpful additions to scar therapy. Heat tolerance must be assessed.
- Extreme caution should be taken if using US. US may be useful in softening connective tissue. Care should be given to avoid overexposure of the epiphyseal plates to US. This exposure can cause premature closure of the epiphyseal plates.

BURNS REQUIRING SPECIAL ATTENTION

Neck

- Correct positioning is essential to avoid neck flexion contracture. No pillows should be used when the patient is lying down.
- In the acute phase, the child should be placed in cervical hyperextension. This can be accomplished by placing a second mattress overlying the first, with the end of the top one slightly above the shoulders. The head then rests on the bottom mattress—do not allow the jaw to drop open.
- The occiput is the prime location for decubiti to occur acutely in children, so the area must be checked on routinely.
- Appropriately fabricated thermoplastic neck conformers do not inhibit function. Soft collars and Philadelphia collars are also used.

Axilla

- Burns of the axilla are often difficult to treat in the pediatric population.
- Of all the burn-related contractures, the axilla is most commonly involved.
- The most successfully used device in an active patient is the airplane splint.

Hands and Feet

- The size of the hands and feet in children combined with a tendency toward rapid contractures in partial and full thickness burns makes this area difficult to treat.
- In hand and foot burns, the development of considerable edema can quickly lead to deformities.
- During the acute phase, fingers and toes should be wrapped separately to maintain average spaces.
- Splinting is universally used to maintain ROM and preserve function.

Splinting Hands and Feet (Table 10–19)

TABLE 10–19 **Splinting of Hands and Feet**

BODY PART	CORRECT SPLINT POSITION
Dorsal hand	MCP flexion 70–90 degrees, IP extension, radial abduction of thumb
Volar hand	MCP/IP full extension, fingers abducted, palmar abduction of thumb
Dorsal foot	Ankle and toes plantar flexion
Sole of foot	Ankle dorsiflexion, toes neutral

IP, interphalangeal; MCP, metacarpophalangeal.
Source: From Molnar GE, Alexander MA. *Pediatric Rehabilitation*. 3rd ed. Hanley & Belfus; 1999, with permission.

PEDIATRIC CANCERS

- Neoplastic disease is the third leading cause of death in children and adolescents in the United States. Solid tumors represent 70% of cases, while acute leukemia represents the remaining 30% (Figure 10–18).
- In children, cancer is the number one cause of death by disease.
- The overall survival rate for children aged 0 to 19 with cancer is about 85%.
- The average age of death of a child with cancer is under 11 years old.

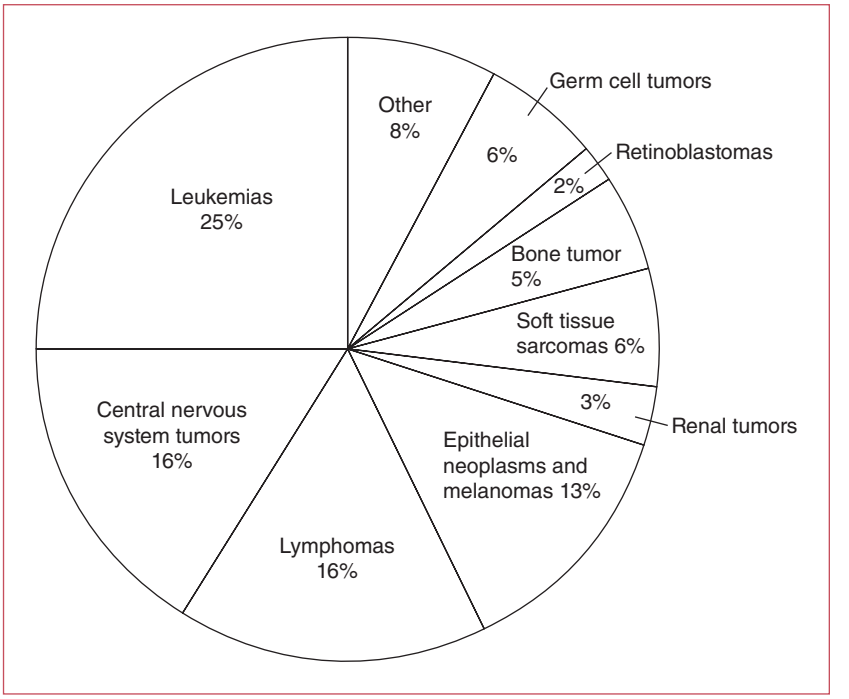


FIGURE 10–18 Major cancers in children <15 years old (approximate incidence).

SOLID TUMORS (70% OF ALL NEOPLASTIC DISEASE IN CHILDREN)

Brain Tumors

- Brain tumors are the second most common type of childhood cancer, with leukemia being the most common. About 50% arise in the posterior fossa.
- Embryonal tumors are the most common malignant CNS tumors in children, of which medulloblastoma accounts for 90%. Medulloblastoma most commonly arises in the midline cerebellar vermis at a median age of 5 to 7 years old in males > females. Patients present with signs of increased intracranial pressure (i.e., headache, nausea, vomiting, mental status changes, and hypertension) and cerebellar dysfunction (i.e., ataxia, poor balance, and dysmetria).
- Cerebellar and fourth ventricle tumors also present with signs of increased intracranial pressure and cerebellar signs.
- Supratentorial tumors present with increased intracranial pressure along with seizures in 40% of cases.
- Brainstem tumors often result in cranial nerve palsies along with hemiparesis and ataxia.

Hodgkin's Disease

- Hodgkin's disease occurs two times more frequently in males than females, with a peak incidence in the third decade, although it has been diagnosed in children as young as 3 years of age.

- Painless cervical adenopathy is the most common presentation in children.
- Etiology is not known.

Non-Hodgkin's Lymphomas

- Non-Hodgkin's lymphoma (NHL) represents a diverse group of hematologic cancers, which include lymphomas other than Hodgkin's lymphoma. Lymphoma is a type of cancer derived from lymphocytes.
- Burkitt's lymphoma (African lymphoma) is a nonlymphoblastic lymphoma found principally in Africa. A viral cause has been proposed, with the Epstein–Barr virus (EBV) suggested. The tumor is characterized by:
  - Predilection for facial bones and mandible
  - Primary involvement of abdominal nodes and viscera
  - Massive proliferation of primitive lymphoid cells

Neuroblastoma

- This tumor arises from cells in the sympathetic ganglia and adrenal medulla.
- It is the third most common pediatric neoplasm in children <5 years old, with the highest incidence at 2 years old.
- Presentation in the abdominal area is commonly associated with distant metastasis and carries a poor prognosis.

Wilm's Tumor (Nephroblastoma)

- In early childhood, the most common abdominal masses encountered are hydronephrosis, neuroblastoma, and Wilm's tumor.
- It develops within the renal parenchyma and enlarges with distortion and invasion of adjacent renal tissue.
- It occurs most often in children 2 to 5 years of age.
- It occurs bilaterally in up to 5% and may be associated with congenital anomalies.

Soft Tissue Tumors

- Rhabdomyosarcoma is the most common malignant soft tissue tumor in children.
- It is associated with neurofibromatosis and most commonly occurs in the head and neck.
- Biopsy is necessary to distinguish this from other soft tissue sarcomas, lymphomas, and neuroblastomas.

Bone Tumors

- In contrast to adults, it is uncommon for bone metastasis to occur in children.
- Benign bone and cartilage tumors include osteochondroma, unicameral bone cyst, osteoid osteoma, eosinophilic granuloma, chondroblastoma, chondromyxoid fibroma, and fibrous dysplasia.
- 🕮 Malignant bone tumors: Osteosarcoma followed by Ewing's sarcoma are the most common types of malignant bone tumors in children and occur more often in children >10 years of age than younger groups:
  - Osteosarcoma typically arises in the metaphysis of long bones. The most common site is the distal femur, followed by the proximal tibia and proximal humerus.
  - Ewing's sarcoma arises in long and flat bones, including the pelvis and most typically in the diaphysis.
- **Figure 10–19** portrays each bone tumor and its characteristic location in long bone. Note that the sites are not exclusive to the femur, which is being used as an example.

Retinoblastoma

- Retinoblastoma is a malignant ocular tumor that occurs at <5 years of age in >90% of cases.
- The tumor may arise sporadically or be inherited.
- Inherited retinoblastomas are often bilateral, whereas the noninherited sporadic mutation form is unilateral.

Germ Cell Tumors

- These tumors are derived from primordial germ cells and can be benign or malignant, gonadal or extragonadal.
- Extragonadal midline sites are involved in about two-thirds of cases, including the sacrococcygeal area, mediastinum, retroperitoneum, and CNS.

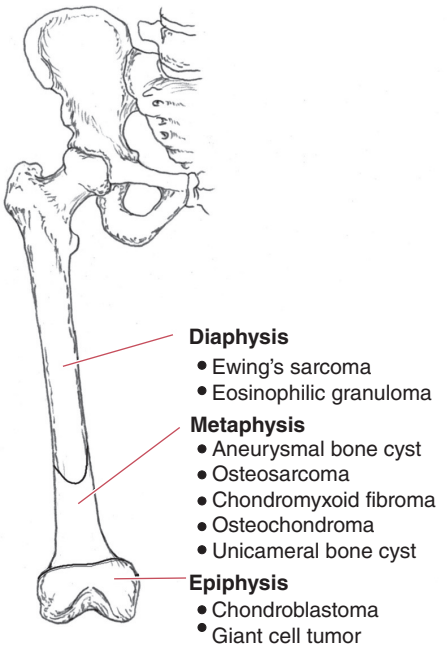


FIGURE 10–19 Characteristic tumor sites in the long bone (not exclusive to the femur, which is illustrated).

LEUKEMIAS (30% OF ALL PEDIATRIC NEOPLASMS)

- 97.5% of leukemias in childhood are acute.
- Acute lymphoblastic leukemia (ALL) is the most common type, accounting for 80% of cases.
- The highest incidence of leukemia occurs between 2 and 5 years of age.
- There is an increased risk of leukemia in patients with chromosomal abnormalities or immune deficiency states.

PEDIATRIC TRAUMATIC BRAIN INJURY

EPIDEMIOLOGY

- Traumatic brain injury (TBI) is the leading cause of death in the United States in children >1 year old and the leading cause of childhood morbidity.
- Between 1.1 and 1.9 million sports- or recreation-related concussions occur annually in children less than 18 years of age.
- In 2013, there were approximately 640,000 TBI-related ED visits among children 0 to 14 years old.
- In 2014, the Centers for Disease Control and Prevention (CDC) reported that more than 23,000 children were hospitalized following a TBI.
- In 2014, 2,529 TBI-related deaths were identified among children.
- Leading causes: Unintentional falls, being struck against or by an object, motor vehicle crashes, sports and recreational activities, other.
  1. For children less than 10 years old, the leading cause of TBI requiring hospitalization is falls, then motor vehicle accident/transport-related injuries.
  2. The order is reversed for children 10 years and older.
- Assaults account for 9% of TBIs requiring hospitalization in children less than 5 years old and 10% in children 15 years of age and older.
- Firearms account for more than a third of fatal injuries from TBI in children 15 years of age and older.

- 70% to 90% of TBI-related ED visits are for mild TBIs.
- Children aged 0 to 4 years are at the highest risk for accidental, nonfatal drowning, with a rate of 16 per 100,000.
- The incidence of childhood ischemic stroke ranges from 1.2 to 7.9 per 100,000 person-years.
  1. The risk of ischemic stroke is highest during the first year of life.

MECHANISM OF INJURY

Primary Injuries

- These occur due to direct impact or the initial deceleration and shearing forces applied to the brain.
- Shearing injuries also result in damage away from any point of impact and include diffuse axonal injury (DAI) and multiple punctate hemorrhages.
- 🕮 The presence or absence of skull fractures is generally not indicative of the severity of brain injury.
- Contrecoup is a cerebral contusion that occurs distant to the point of impact against an object.

Secondary Injuries

- These occur as a sequelae of the initial insult and contribute to additional injury to the brain.
- Causes include factors that interfere with cerebral perfusion or oxygenation (see also Chapter 2, "Traumatic Brain Injury"):
  - Hypotension
  - Increased intracerebral pressure (ICP) secondary to cerebral edema, acute hydrocephalus, or mass lesion
  - Hypoxia
  - Midline shift or herniation that may lead to infarction because of pressure or traction on cerebral vessels
- In young children, incomplete myelination may result in a greater risk of shearing injury. Their relatively large heads may increase the likelihood of injury secondary to increased rotational forces.
- Nonaccidental injuries are a result of acceleration–deceleration forces and are generally associated with retinal hemorrhages, fractures (spiral fractures or at various stages of healing), and multiple injuries.

Associated Injuries

- Because a large number of TBIs in children are due to motor vehicle accidents or other high-speed accidents, almost one-half of all children who sustain a TBI also suffer other injuries as well.
- The associated injuries can affect long-term outcome and complicate acute management.
- Other injuries sustained include:
  - Spinal cord injury in 5% to 10% of TBI children
  - Brachial plexus secondary to traction injury (see the following box).
  - Fractures
  - Perforated viscus
  - Liver and splenic lacerations

Neonatal Brachial Plexus Injuries

- 🕮 Brachial plexus injuries are typically due to:
  - Trauma
  - Obstetrical complications
- 🕮 **Upper trunk brachial plexopathy (Erb–Duchenne Palsy)**
  - Due to sudden traction to the neck, causing injury to the upper trunk of the brachial plexus (C5–C6 roots).

(*continued*)

- 🕮 **Lower trunk brachial plexopathy (Klumpke's Palsy)**
  - Due to violent upward pull of the shoulder, causing damage to the lower trunk (C8–T1 roots). Horner's syndrome can be associated with injury of the C8 and T1 roots, which affects the superior cervical sympathetic ganglion.
  - Klumpke's paralysis is rare in the setting of traumatic birth palsy, as it typically results from a fall onto a hyperabducted shoulder, penetrating trauma, or a mass lesion (e.g., tumor).
- **Entire brachial plexus injury**: Secondary to injuries

SEVERITY OF BRAIN INJURY

- The Pediatric Glasgow Coma Scale (GCS) is usually determined within hours of injury.
- (Table 10–20 and 10–21):
  - GCS 13 to 15 is considered a mild brain injury.
  - GCS 9 to 12 is considered a moderate brain injury.
  - GCS 3 to 8 is classified as a severe brain injury, and the patient is considered to be comatose.
- Adaptations of the GCS for verbal response have been developed for children ≤2 years old (see Table 10–21).

🕮 TABLE 10–20 Pediatric Glasgow Coma Scale

EYE OPENING RESPONSE	
Spontaneous	4
To speech	3
To pain	2
None	1
VERBAL RESPONSE: CHILD (INFANT MODIFICATION)	
Oriented (coos, babbles)	5
Confused conversation (irritable cry, consolable)	4
Inappropriate words (cries to pain)	3
Incomprehensible sounds (moans to pain)	2
None	1
BEST UPPER LIMB MOTOR RESPONSE: CHILD (INFANT MODIFICATION)	
Obeys commands (normal movements)	6
Localizes pain (withdraws to touch)	5
Withdraws to pain	4
Flexion to pain	3
Extension to pain	2
None	1

Source: Adapted from Mandt MJ, Faries G. Emergencies & Injuries. In: Hay WW, Jr., Levin MJ, Sondheimer JM, Deterding RR, eds. *CURRENT Diagnosis & Treatment: Pediatrics*. 19th ed. McGraw-Hill Professional; 2008:294–312, with permission.

TABLE 10–21 Glasgow Coma Scale for Young Children: Modification of Scoring Verbal Responses

VERBAL SCORE	ADULT AND OLDER CHILDREN	YOUNG CHILD (≤2 YEARS OLD)
5	Oriented	Smiles, oriented to sound, follows objects, interacts
4	Confused, disoriented	Cries but consolable, interacts inappropriately
3	Inappropriate words	Cries but is inconsistently consolable, moaning
2	Incomprehensible sounds	Inconsolable crying, irritable
1	No response	No response

Source: From Alexander MA, Matthews DJ, eds. *Pediatric Rehabilitation: Principles and Practice*. Demos Medical Publishing; 1999.

TABLE 10–22 Brain Injury Severity Rating by Duration of Consciousness

	MILD	MODERATE	SEVERE	PROFOUND
Initial Glasgow Coma Scale	13–15 with no deterioration	9–12 with no deterioration	3–8	
Posttraumatic amnesia	<1 hour	1–24 hours	>24 hours	
Duration of unconsciousness	<15–30 minutes	15 minutes–24 hours	1–90 days	>90 days

Source: From Alexander MA, Matthews DJ, eds. *Pediatric Rehabilitation: Principles and Practice*. 4th ed. Demos Medical Publishing; 2010, with permission.

COMMON MOTOR DEFICITS

- Focal damage may result in hemiparesis.
- Diffuse damage: Deficit in balance, coordination, and initiation. Overall, 79% achieve independence in mobility.
- Balance deficits: Impairment in cochlear and vestibular functions
- Tremor
- Dystonia: More common in children than in adults status post-TBI
- Spasticity/rigidity, combined spasticity/ataxia

COMMON SENSORY DEFICITS

- Anosmia: Impaired olfaction, usually secondary to damage to olfactory bulbs and tracts, temporal lobes, or subfrontal areas.
- Hearing impairment: May be due to central processing deficit, peripheral nerve damage, cochlear injury, or disruption of the middle ear structures. Injuries to cranial nerve VIII are frequently associated with basilar skull fracture.
- Visual impairment: Injuries to cranial nerves, eyes, optic chiasm, tracts, radiations, or cortical structures. Optic nerve injury occurs in 1.5% of cases.

COGNITIVE DEFICITS

- Cognitive and communication deficits are believed to be the largest cause of disability in TBI.

- Impairment of arousal and attention: Experimental evidence exists that neurotransmitters decrease after brain injury. Trial of dopamine, norepinephrine, tricyclic antidepressants, and serotonin may be beneficial.
- Hyperactive children are more likely to sustain TBI. One study revealed 35% had a history of learning disability, attention deficit, or emotional problems before the accident.
- Agitation: Damage to the frontal lobes and subcortical areas may result in agitation.
- Memory impairment: Particularly affected in moderate to severe injuries. Academic performance is directly correlated to the severity of injury. Majority of improvement in IQ occurs in the first 4 months after TBI.
- Communication impairment: Two-thirds present with difficulty in communication, including naming, verbal fluency, and expression. One-third present with dysarthria. The age at injury may influence language outcome, as the young child has experienced less language development before injury.
- Behavioral sequelae: Deficits in impulse control and disinhibition may relate to frontal lobe injury. Adolescents may exhibit hypersexuality.
- Abnormal emotional expression: Initial lack of emotional expression may present as long-term emotional lability.
- Impairment in abstract reasoning
- Also associated egocentricity
- Social isolation: Difficulty forming lasting social relationships

MEDICAL PROBLEMS ASSOCIATED WITH TRAUMATIC BRAIN INJURY

Neuroendocrine Dysfunction

- Diabetes insipidus (DI): Excessive water loss secondary to deficiency of antidiuretic hormone (ADH), which is produced in the hypothalamus. Acute-onset DI may be a poor prognostic factor for post-TBI survival.
- Syndrome of inappropriate ADH (SIADH): Characterized by decrease in urinary output, hyponatremia, and decreased serum osmolarity.
- 🕮 Hypopituitarism: Presents with growth failure and delayed or arrested puberty. Patients present with deficiencies in production of all pituitary hormones, with growth hormone, luteinizing hormone, and follicle-stimulating hormone being most deficient (Acerini et al., 2006).
- Pituitary deficiency prevalence ranges from 5% to 57%.
- Cerebral salt wasting (CSW): Direct neural effect on renal tubular function
- Precocious puberty: Initial signs may present 2 to 17 months after TBI. Girls more affected than boys (54.5% vs. 4.5%). Clinically show precocious secondary sexual development, accelerated linear growth, advanced bone ages >2 years old, and shortened adult stature secondary to premature epiphyseal closure.

Respiratory Dysfunction

- Acutely intubated as part of the initial management.
- Pneumonia may be an early respiratory complication.
- Prolonged intubation can result in tracheal stenosis in the glottic area, tracheomalacia, and vocal cord injury/paralysis.

Gastrointestinal Concerns

- Nutritional concerns are that the TBI child is hypermetabolic.
- Tube feedings are used because of decreased level of responsiveness.
- Gastroesophageal reflux should be assessed before a gastrostomy tube is placed.

Bowel Management

- It is important to establish routine bowel-management programs.
- Early after injury, bowel motility decreases on the basis of the injury itself or because of narcotics.

Bladder Management

- Acutely, short-term bladder management is recommended to ensure that fluid intake and output are balanced.
- Long-term bladder management becomes a more cognitively based activity.
- If incontinence persists, the patient may have a neurogenic bladder. Clinical evaluation and treatment should then be performed.

Central Autonomic Dysfunction

- Clinical entity defined as symptoms of unexplained hyperthermia, systemic hypertension, diaphoresis, generalized rigidity, decerebrate posturing, and tachypnea after TBI.
- Occurs in 14% of children with severe TBI.
- The mechanism is felt to involve hypothalamic or brainstem dysfunction.
- Presence of central autonomic dysfunction after TBI is correlated with a more protracted period of unconsciousness and worse cognitive and motor outcomes >1 year after injury.

Heterotopic Ossification

- Ectopic bone formation occurs in 14% to 23% of pediatric TBI patients.
- More common in children >11 years old, in those with more severe injury, and in those who had two or more extremity fractures.
- It most commonly affects the hips and knees, and presents with pain, decreased ROM, and sometimes swelling.
- Heterotopic ossification (HO) is usually diagnosed a month or later after injury.
- It is associated with poor outcome.
- Deep venous thrombosis is an unusual complication in children but may be seen in association with HO.
- Treatment includes gentle ROM, splinting and positioning, and NSAIDs. Do not use etidronate because it has been reported to result in a reversible rachitic syndrome in growing children.

Posttraumatic Epilepsy

- The incidence of both early (within first week) and late (after 1–2 weeks) seizures is increased after TBI.
- Patients with two or more late seizures after TBI are considered to have posttraumatic epilepsy (PTE). The risk of development of PTE is correlated with severity of TBI:
  - Incidence of PTE was 22% across all severities of TBI.
- Prophylaxis with antiepileptic medication is not recommended. There has been no reported efficacy in prevention of PTE using phenytoin therapy.

Cerebral Atrophy and Posttraumatic Hydrocephalus

- Enlargement of the ventricular system is commonly seen after severe TBI in children.
- This results from cerebral atrophy (hydrocephalus ex vacuo) or obstruction of cerebrospinal fluid (CSF) flow (hydrocephalus).
- Cerebral atrophy is more frequently seen after severe brain injury than true hydrocephalus.

SURVIVAL

- More than two-thirds of deaths from brain injury occur at the scene or en route to the hospital.
- For those children with significant injury, up to 47% of hospital costs are due to inpatient rehabilitation. Most children are discharged to home after TBI. Generally, all children who had even minimal responsiveness survive for years.
- Death in children with profound brain injuries is seen more commonly in those who remained in vegetative states >90 days after an anoxic or traumatic injury compared with adults:
  - In adults, 50% of patients in vegetative states die within 1 year of injury, while 50% of children who were in vegetative states at 1 year after injury were still living 7 to 8 years later.

LONG-TERM IMPAIRMENT

- Children with minor TBI are clinically indistinguishable from age-matched controls at 1 year postinjury. These children rarely have impairments that can be attributed to the accident.
- In severe TBI, 87% of children unconscious for more than 6 hours had a good recovery and were able to lead fully independent lives with or without minimal neurologic deficit. 73% became independent in ambulation and self-care within a year of injury.
- Children with profound brain injury, who were unconscious for more than 90 days, have a less favorable prognosis for recovery.
- 🕮 In general, a traumatic cause of brain injury has a much better prognosis than an anoxic etiology. 24% of children with TBI who are unconscious at 1 month postinjury regain awareness after 3 months, whereas only 11% of those with anoxic injury did so. After 1 year postinjury, 81% of children with TBI were alive, with only 29% remaining unconscious, while only 75% of children with anoxic injury were alive, with 65% remaining unconscious (Mayfield et al., 2009).
- 75% of children with TBI who are unconscious for more than 90 days eventually regained consciousness, whereas only 25% of those with anoxic injury did so. In general, a traumatic cause of brain injury has a better prognosis than an anoxic etiology.
- 61.6% of children with moderate-to-severe TBI require specialized medical and educational services, compared with 14.3% of children with mild TBI.
- Survival and neurologic outcome are worse for abused children than for other causes of TBI.
- Final outcome of children with diffuse TBI is worse in those youngest at age of injury.
- Children who sustain a TBI before 2 years old exhibit more global neurobehavioral impairments than children who sustain a TBI after 7 years of age.

CEREBRAL PALSY

DEFINITION OF CEREBRAL PALSY

- 🕮 Cerebral palsy (CP) is a disorder primarily of movement control and posture but is also associated with cognitive and sensory-associated problems resulting from a nonprogressive lesion to an immature brain.
- Can occur in utero (prenatal period), near time of delivery (perinatal period), or within the first 3 years of life (postnatal period).
- 🕮 CP is the leading cause of childhood disability with an incidence of 2 to 3 per 1,000 births.
- There is evidence for genetic predisposition to the development of CP.

RISK FACTORS FOR CEREBRAL PALSY: PRENATAL, PERINATAL, AND POSTNATAL (TABLE 10–23)

TABLE 10–23 Risk Factors for Cerebral Palsy

PRENATAL RISK FACTORS	PERINATAL RISK FACTORS	POSTNATAL RISK FACTORS
🕮 Majority of CP cases (70%–80%) occur during the prenatal period. Risk factors include the following: • **Prenatal intracranial hemorrhage:** – Outcome depends on degree of associated parenchymal injury. • **Placental complications** • **Gestational toxins:** – Iodine deficiency can lead to diplegia. – Organic mercury intoxication can lead to quadriplegia. • **Gestational teratogenic agents** • **Congenital malformations of the brain and cerebral vascular occlusions during fetal life**	• **Complications of prematurity** • Birth at <32 weeks of gestation • Low birth weight <2,500 grams • 🕮 Prematurity remains the most common antecedent of CP due to a combination of: – Immaturity – Fragile brain vasculature – Physical stresses of immaturity that predispose children to compromise of cerebral blood flow – Blood vessels in the watershed zone next to the lateral ventricles in the capillaries of the germinal matrix that are particularly vulnerable • **Complications of full-term infant:** – Placenta previa – Placental abruption – Meconium aspiration resulting in neonatal asphyxia	• **Trauma:** – Fall – Child abuse (i.e., shaken baby syndrome—look for retinal hemorrhage) – MVA • **Toxins:** – Heavy metals, especially lead – Organophosphates • **Stroke syndromes with hemiplegia which can result from:** – Sickle-cell anemia – AVM rupture – Congenital heart disease (especially tetralogy of Fallot) • **Infection** – Bacterial – Viral – Meningitis (especially in first 6 months)
• **Acquired congenital infections (TORCH):** – Toxoplasmosis – Rubella – CMV – Herpes • **Maternal causes:** – Seizures – Hyperthyroidism – Mental retardation • **Socioeconomic factors** • **Reproductive inefficiency** • **Prenatal hypoxic-ischemic injury as a result of:** – Idiopathic (most common) – Multiple pregnancies – Maternal bleeding – Maternal drug use Note: Prenatal factors may lead to premature birth or intrauterine growth restriction of both term and preterm infants.	• **Hyperbilirubinemia:** – Rh incompatibility – G6PD deficiency – ABO incompatibility – Results in kernicterus with deposition of bilirubin in the cranial nerve nuclei and basal ganglia, causing subsequent athetoid CP • **Difficult/traumatic delivery** – Typically SDH causing mechanical trauma to the brain at birth can result in CP (spastic hemiplegia type) • **Infection:** – Viral – Bacterial • **Seizures** • **Bradycardia and hypoxia** • **Perinatal intracranial hemorrhage:** – Outcome depends on the degree of associated parenchymal injury	• **Neoplastic disease** • **Anoxia** – i.e., near drowning can cause anoxic encephalopathy • **Intracranial hemorrhage**

AVM, arteriovenous malformation; CMV, cytomegalovirus; CP, cerebral palsy; G6PD, glucose 6 phosphodehydrogenase; MVA, motor vehicle accident; SDH, subdural hematoma; TORCH, toxoplasmosis, other agents such as varicella virus or parvovirus, rubella virus, cytomegalovirus, herpes virus/HIV, syphilis.

CLASSIFICATION OF CEREBRAL PALSY (TABLES 10–24 AND 10–25)

- Current methods try to incorporate a functional basis for classification. CP may manifest itself differently as the child ages. The modified neurologic classification system divides patients into the following categories:
  - Spastic (pyramidal) CP
  - Dyskinetic (extrapyramidal) CP
  - Mixed type (combination) CP

SPASTIC TYPES OF CEREBRAL PALSY (75%)

- Children with spastic CP exhibit significant upper motor neuron (UMN) signs, such as muscle spasticity and hyperreflexia/clonus. Extensor Babinski response (abnormal at >2 years old) may be present in addition to other persistent primitive reflexes.
- The spastic group is divided into five subtypes that are named by the part(s) of the body involved.

1. Spastic Monoplegia

- This type is rarely seen. The child has isolated UE or LE involvement and usually has a mild clinical presentation.

🕮 2. Spastic Diplegia (Most Common Type of Cerebral Palsy)

- Of premature infants that develop CP, 75% are spastic diplegics.
- Primarily LE >> UE involvement with a history of prematurity.
- Infants clinically present with a history of early hypotonia followed by development of spasticity. Developmental delays, commonly in the area of gross motor function, are also noted.
- History of intraventricular hemorrhage is also typical, especially at 28 to 32 weeks of gestation. MRI may show periventricular leukomalacia or posthemorrhagic porencephaly.
- LE spasticity is caused by damage to pyramidal fibers within the internal capsule. Muscles that are affected typically include the hip flexors, adductors, and gastrocnemius muscles. Contractures result secondary to long-term spasticity.
- Diplegic gait pattern includes classic scissoring gait with toe walking.
- Mild coordination problems result in the UEs with UMN findings in the LEs.
- Ocular findings include strabismus in 50% and visual deficits in 63%.
- Seizures occur in 20% to 25% and cognitive impairment found in 30%.

3. Spastic Triplegia

- Involves three extremities with the bilateral LEs and one UE classically affected.
- Spasticity results in the involved limbs with mild coordination deficits in the uninvolved limb.
- Characteristic scissoring and toe walking are observed.

4. Spastic Tetraplegia

- All extremities are involved.
- Pattern of truncal hypotonia with appendicular hypertonia or total body hypertonia exists.
- Often a history of difficult delivery with evidence of perinatal asphyxia.
- Approximately 50% have a prenatal origin, 30% perinatal, and 20% postnatal.
- MRI in the preterm child shows a periventricular leukomalacia.
- Opisthotonic posturing may begin in infancy, often persisting in the severely involved. Oromotor dysfunction, pseudobulbar involvement, and risk of aspiration with feeding difficulties occur and may require feeding tube placement.
- Cognitive involvement results in a large percentage of intellectual disability.
- Seizures occur in 50% of affected children.
- Spasticity and persistent primitive reflexes contribute to making these children the most severely involved of those with CP.

5. Spastic Hemiplegia

- One side of the body is involved, usually the arm more than the leg.
- 70% to 90% are congenital.
- 10% to 30% are acquired secondary to vascular, arteriovenous malformations, inflammatory, or traumatic causes.
- MRI reveals evidence of unilateral lesions in 66% of cases.
- In term infants, the cause is usually secondary to prenatal events.
- In premature infants, asymmetric periventricular leukomalacia is a common cause.
- Hemiparesis is usually evident by 4 to 6 months of age, with hypotonia being the first indicator. Other indicators include premature preferential hand dominance.
- There is a slightly higher incidence of right-sided involvement.
- Average age of walking is 24 months.
- Cranial nerves may be involved (facial weakness).
- Growth retardation of the affected side is often seen with associated spasticity.
- Sensory deficits on the ipsilateral side occur in 68%.
- Visual deficits occur in 25% of hemiplegics, cognitive impairment in 28%, and seizures in 33%.
- Perceptual motor deficits are common, causing learning disabilities.

🕮 DYSKINETIC TYPES OF CEREBRAL PALSY

- Dyskinesias are characterized by abnormal extrapyramidal movement patterns secondary to dysregulation of tone, defects in postural control, and coordination deficits.
- **Athetosis:** Slow writhing involuntary movements, particularly in the distal extremities.
- **Chorea:** Abrupt, irregular jerky movements, usually occurring in the head, neck, and extremities.
- **Choreoathetoid:** Combination of athetosis and choreiform movements. They are generally large amplitude involuntary movements. The dominating pattern is the athetoid movement.
- **Dystonia:** A slow rhythmic movement with tone changes generally found in the trunk and extremities; associated with abnormal posturing.
- **Ataxia:** Uncoordinated movements often associated with nystagmus, dysmetria, and a wide-based gait:
  - Classic movement patterns emerge sometime between 1 and 3 years of age. Severely affected children have persistent hypotonia. Movement patterns typically increase with stress or purposeful activity. During sleep, muscle tone is normal, and involuntary movement stops.
  - Pseudobulbar involvement presents with dysarthria, dysphasia, drooling, and oromotor dyskinesias. These children have normal intelligence 78% of the time.
  - 🕮 There is a high incidence of sensorineural hearing loss, which has been associated with hyperbilirubinemia and neonatal jaundice.

MIXED TYPES OF CEREBRAL PALSY

- Mixed types of CP exhibit patterns of both spastic and dyskinetic types of CP. The most common mixed type is spastic athetoid (predominant dyskinetic movement pattern with an underlying component of spasticity).
- The modified neurologic classification system divides patients into the following categories.

TABLE 10–24 Classification of Cerebral Palsy

TYPE	SPASTIC (PYRAMIDAL) CP	DYSKINETIC (EXTRAPYRAMIDAL) CP	MIXED TYPES
% of CP	*75%*	*25% (Both Dyskinetic and Mixed Types)*	
Clinical Signs	Manifest signs of upper motor neuron involvement: Hyperreflexia Clonus Extensor Babinski response (abnormal at >2 years) Persistent primitive reflexes Overflow reflexes 📖 **Subgroups:** **Spastic monoplegia** (controversial; usually a mild hemiplegia) **Spastic diplegia** (most common type of CP) **Spastic triplegia** **Spastic tetraplegia** **Spastic hemiplegia**	Dyskinesias are characterized by extrapyramidal movement patterns secondary to abnormal regulations of tone, defects in postural control, and coordination deficits: **Athetosis**—slow writhing, involuntary movements, especially in the distal extremities. **Chorea**—abrupt irregular jerky movements, usually occurring in the head, neck, and extremities. **Choreoathetoid**—combination of athetosis and choreiform movements; the dominating pattern is the athetoid movement with intermittent large-amplitude involuntary movements. **Dystonia**—a slow rhythmic movement with tone changes generally found in the trunk and extremities, associated with abnormal posturing. **Ataxia**—unsteadiness with uncoordinated movements associated with nystagmus, dysmetria, and a wide-based gait.	Exhibit pattern of both spastic and dyskinetic types of CP. Example—spastic/athetoid—predominantly dyskinetic movement pattern with underlying component of spasticity.

CP, cerebral palsy.

TABLE 10–25 Major Categories of CP: Spastic, Dyskinetic, and Mixed Types

SPASTIC TYPE (75% OF ALL CASES OF CP) BASED ON TOPOGRAPHIC DISTRIBUTION OF SPASTICITY			
TYPE	**ETIOLOGY**	**WILL THEY WALK?**	**ASSOCIATED FINDINGS**
Hemiplegic	• **Most cases congenital (some acquired 10%–30%)** • **Focal perinatal injury** • Highest incidence of CT/MRI abnormalities in distribution of MCA • **Infarction in a vascular distribution** (usually MCA) results in focal and multifocal ischemic brain necrosis	• Ambulation usually achieved by 2 years of age (unless severe retardation is associated)	• Most common presentation: Failure to use the involved hand (i.e., preferential hand used prior to 1 year of age) • Arm more involved than leg • Speech is preserved—children have the ability to switch dominance of hemisphere up until approximately age 6 • Asymmetric crawl seen • Refractory error; may have hemianopsia • Initial seizures may occur as late as 5 years of age • Mild intellectual disability or no intellectual disability • Cortical sensory deficit

(*continued*)

TABLE 10–25 Major Categories of CP: Spastic, Dyskinetic, and Mixed Types (*continued*)

SPASTIC TYPE (75% OF ALL CASES OF CP) BASED ON TOPOGRAPHIC DISTRIBUTION OF SPASTICITY			
TYPE	**ETIOLOGY**	**WILL THEY WALK?**	**ASSOCIATED FINDINGS**
Diplegic (Little's disease) 🕮 Most common type of CP seen in premature infants	• **Ischemia** due to hypoperfusion of the germinal matrix of the periventricular region of the premature fetus • The periventricular white matter resides within the border zones between the penetrating branches of the major vessels and is exquisitely sensitive to decreases in cerebral perfusion • Result is PVL. This region of white matter is traversed by the descending fibers of the motor cortex. In the corona radiata, descending fibers from the motor cortex are arranged with those subserving the LE medially and UE laterally. • Therefore, smaller lesions result in diplegic CP (i.e., involving mainly the LE). (Note: Larger lesions affect both the UE and LE and result in quadriplegic CP as noted in the text that follows.)	• Most diplegics ambulate—some require assistive devices	• Disproportionate involvement of the legs, although upper extremity motor perceptual dysfunction also seen • Developmental delay in gross motor skills • Ocular findings include strabismus in 50% of the cases (visual deficits in 63%) • Mild cognitive impairment (30%) or no intellectual disability • Seizures occur in 25% • UMN findings in the lower extremities • Initial period of hypotonicity; later these patients develop increased tone in the lower extremities
Tetraplegic 🕮 This group has the highest incidence of significant disability: • 25% severely involved • 50% moderately involved	• **Major hypoxic event** (perinatal asphyxia) usually a history of difficult delivery • Result is a more severe degree of ischemia (more than diplegic CP) with more advanced PVL. This larger lesion results in UE and LE involvement (quadriplegic CP) • **Parasagittal cerebral injury**—*injury to bilateral cortical zones,* causing necrosis within the border watershed zones. Injury to this region affects the medial motor cortex, which affects UE more severely than LE (think of the homunculus) • **Focal and multifocal ischemic brain lesions**	• One-fourth are independent in ambulation, modified ADL • One-half require assisted ambulation, assisted ADL • One-fourth are completely disabled	• Strabismus • Legs are usually more involved than arms—asymmetries not unusual • Intellectual disability can be significant • Oromotor dysfunction, pseudobulbar involvement present risk of aspiration • Feeding difficulties—may need G-tube • UMN signs in all limbs • One-half have seizures • Must be monitored for hip dislocation and scoliosis • Initial period of hypotonia, which eventually develops into extensor spasticity • The more persistent the tone, the poorer the prognosis. • Opisthotonus and precocious head raising can be an early manifestation of extensor hypertonicity.

(*continued*)

TABLE 10–25 Major Categories of CP: Spastic, Dyskinetic, and Mixed Types (*continued*)

SPASTIC TYPE (75% OF ALL CASES OF CP) BASED ON TOPOGRAPHIC DISTRIBUTION OF SPASTICITY			
TYPE	**ETIOLOGY**	**WILL THEY WALK?**	**ASSOCIATED FINDINGS**
DYSKINETIC AND MIXED TYPES (COMPRISE 25% OF ALL CASES OF CP)			
Dyskinetic Dyskinetic disorders are characterized by abnormal extrapyramidal movement patterns. (Dyskinetic movements are defined previously in the modified neurologic classification system.)	• **In the past, most cases were associated with** kernicterus (neonatal hyperbilirubinemia causing bilirubin deposition in the basal ganglia). May be due to hemolysis secondary to Rh or ABO blood type incompatibility or hematoma breakdown. • Diffuse anoxia resulting in hypoxia of the basal ganglia and thalamus	• One-half of children attain walking, most of them after 3 years of age • UE function is adequate for ADLs • One-half of children are nonambulatory, dependent in ADLs	• Athetosis—slow writhing, involuntary movements especially in the distal extremities • Seizures in 25% • 🕮 Sensorineural deafness (high incidence) • Paralysis of upward gaze • These children are generally hypotonic at birth. • Classic movement patterns emerge some time between 1 and 3 years of age. • The child develops involuntary movements first in the hands and fingers. Abnormal movements are usually evident in all extremities by 18 months to 3 years of age. • Pseudobulbar signs are present with drooling, oromotor dyskinesias, and dysarthria. Muscle tone is normal during sleep. • DTR is normal to slightly increased. • Tension athetosis when moving the limb increases tone. • Children have normal intelligence 78% of the time. Nonambulatory patients are at risk for hip dysplasia and scoliosis. • The UE is often more involved than the LE.
Mixed types Include descriptions from both spastic and dyskinetic classifications, i.e., spastic athetoid	• Mixed etiology	• Dependent on classification	• Ocular findings: Kernicterus causes paralysis of upward gaze (rarely seen). Nystagmus is present in the ataxic type.

ADLs, activities of daily living; CP, cerebral palsy; DTR, deep tendon reflex; LE, lower extremity; MCA, middle cerebral artery; PVL, periventricular leukomalacia; UE, upper extremity; UMN, upper motor neuron.

GROSS MOTOR FUNCTION CLASSIFICATION SYSTEM FOR CEREBRAL PALSY

- Gross motor function classification system (GMFCS) is a functionally based system to standardize gross motor function in the CP child.
- **Level 1**: Walks without restrictions; limitations in more advanced gross motor skills.
- **Level 2**: Walks without assistive devices; limitations with walking outdoors and in the community.
- **Level 3**: Walks with assistive mobility devices; limitations with walking outdoors and in the community.

- **Level 4**: Self-mobility with limitations; transported or use power mobility outdoors and in the community.
- **Level 5**: Self-mobility severely limited even with use of assistive devices.

TYPICAL GAIT ABNORMALITIES IN CEREBRAL PALSY

Spastic Diplegia

- Scissoring gait pattern:
  - Hips flexed and adducted
  - Knees flexed with valgus
- Ankles in equinus (toe walking)

Spastic Hemiplegia

- Weak hip flexion and ankle dorsiflexion
- Overactive posterior tibialis
- Hip hiking or hip circumduction
- Supinated foot in stance phase
- UE posturing

Crouch

- Tight hip flexors
- Tight hamstrings
- Weak quadriceps
- Excessive dorsiflexion in both diplegics and quadriplegics

🕮 WILL MY CHILD WALK? (TABLE 10–25)

- This is usually the most frequent question asked by the parent of a newly diagnosed CP child. Several factors are relevant. The best indicator of how the child will do is how the child is doing.
- 🕮 **Sitting**: Alexander (2010) has shown that if independent sitting occurs by age 2, prognosis for ambulation is good.
- **Crawl**: Badell felt that ability to crawl on hands and knees by 1.5 to 2.5 years is a good prognostic sign (Badell-Ribera, 1985).
- **Primitive reflexes**: Persistence of three or more primitive reflexes at 18 to 24 months is a poor prognostic sign (**Table 10–28**).
- Prognosis also depends on the type of CP (**Table 10–25**).

ASSOCIATED DEFICITS IN CEREBRAL PALSY (TABLE 10–26)

Intellectual Disability

- The incidence of associated disabilities in CP varies. The overall incidence of intellectual disability is up to 50%.
- Microcephaly, seizures, and severe neuromuscular dysfunction are associated with increased risk of intellectual deficit.
- Spastic tetraplegic has the highest rate of intellectual disability, while spastic hemiplegia and diplegia have the lowest.

Seizures

- The overall incidence of seizure in children with CP is up to 50%. Seizures are more common in spastic tetraplegics (50%) and less frequent in diplegics and dyskinetics (25%–33%). Grand mal with tonic–clonic convulsions is a common type.

TABLE 10–26 Associated Deficits in Cerebral Palsy

Intellectual disability	50% incidence, most common in severely spastic tetraplegia
Seizures	50% incidence, most frequent in hemiplegia and spastic tetraplegia
Oromotor	Difficulty sucking, swallowing, and chewing; poor lip closure, tongue thrust, drooling, dysarthria; most common in spastic quadriplegia, dyskinetic
Gastrointestinal	Reflux, constipation
Dental	Enamel dysgenesis, malocclusion, caries, gingival hyperplasia (secondary to phenytoin)
Visual	Strabismus, refractory errors; hemianopsia in hemiplegia
Hearing impairment	Infection (TORCH), medications, kernicterus
Cortical sensory deficit	Hemiplegia
Pulmonary	Deficient ventilation, bronchopulmonary dysplasia in premature infants; microaspirations with oromotor dysfunction
Bowel and bladder	Dysfunction of central neuromotor control and cognitive developmental status of the child
Behavior	Attention deficit disorder, distractibility, impulse control, overt hyperkinesis, organic pseudobulbar palsy
Language disorders	Developmental, pseudobulbar supranuclear spastic paralysis, incoordination of muscles innervated by the lower cranial nerves

TORCH, toxoplasmosis, other agents such as varicella virus or parvovirus, rubella virus, cytomegalovirus, herpes virus/HIV, syphilis.
Source: From Molnar GE, Alexander MA. *Pediatric Rehabilitation*. 3rd ed. Hanley & Belfus; 1999, with permission.

Visual Deficits

- Deficits of extraocular movements and vision are also common in CP.
- Prevalence is 30% to 100%, with 6% to 11% functionally blind.
- Strabismus is the most frequent visual deficit at 25% to 60% of all cases, with the highest rate in spastic diplegics and tetraplegics.
- Esotropia is more frequent than exotropia.
- Paralysis of conjugate upward gaze is a clinical manifestation of kernicterus.
- Nystagmus is present in the ataxic type.
- A homonymous hemianopsia occurs in hemiparetic CP in 25% of cases.
- Retinopathy of prematurity occurs in preterm infants.

Hearing Impairment

- The characteristic hearing loss in CP is a sensorineural impairment that occurs in up to 12% of CP children <15 years old, with a fourfold increased prevalence in athetosis than in spasticity.
- Kernicterus is the most common cause of sensorineural hearing loss in athetosis. Other causes include intrauterine infections, especially rubella, cytomegalovirus, toxoplasmosis, and syphilis, as well as perinatal hypoxia, meningitis, encephalitis, and ototoxic drugs.

Language Disorders

- Developmental language disorders of verbal and written communication also occur. After 1.5 to 2 years of age, insults to the dominant hemisphere lead to aphasia. Although most children show significant recovery from aphasia acquired before 8 to 10 years, they rarely regain premorbid levels.

- Defective speech results from pseudobulbar palsy and supranuclear spastic paralysis or dyskinetic incoordination of the muscles innervated by the lower cranial nerves. Most athetoids and 50% of bilateral spastics have some dysarthria.
- 🕮 Speaking in two- to three-word sentences by age 3 is a good indication of intellectual potential.
- 31% to 88% have communication disorders.

Respiratory Impairment

- Respiratory impairment may also occur in CP children. Decreased vital capacity and aerobic working capacity is seen both in spastic and athetoid types. Restrictive pulmonary disease accompanies scoliosis. Chronic aspiration leads to interstitial fibrosis.

Behavioral Disorders

- Behavioral and emotional disorders occur in 25% of children with CP.
- Disorders of behavior may present with attention deficit, distractibility, disturbances of impulse control, and overt hyperkinesis.
- Behavior disorders also include true emotional lability as part of an organic pseudobulbar palsy consisting of dysarthria, drooling, and poor chewing.
- Poor peer acceptance can lead to a negative self-image; school problems, depression, and anger may be exacerbated during normal periods of transition, that is, preschool to kindergarten and in early adolescence.
- The more mildly physically involved child may have more difficulty and need more psychosocial support.

Gastrointestinal Problems

- Symptoms are frequent. Gastroesophageal reflux often requires medical management. Constipation is exaggerated by immobility and abnormal diet and fluid intake.

Bowel and Bladder Dysfunction

- Management is usually related to dysfunction of the central neuromotor control and the cognitive development status of the child.

Oromotor Problems

- 27% to 90% have difficulty with eating or drinking.
- These may lead to difficulty swallowing, sucking, and chewing. Motor incoordination is manifested by poor lip closure, retraction or thrusting of the tongue, and decreased tongue movements.
- Feeding difficulties can contribute to malnutrition and aspiration. Dysphagia evaluation, modified barium swallow, and fiber optic endoscopy evaluation may be needed. A percutaneous gastric tube may be necessary in certain cases.

Dental Problems

- Issues include malocclusion, enamel dysgenesis secondary to palatal distortions, and abnormal oromotor reflexes.
- CP children are also at increased risk for cavities due to poor handling of secretions and food, as well as chronic drooling. Medications, such as scopolamine patches, can be used to address the problem of drooling.

PROGNOSIS

- About 90% of children with CP survive to adulthood. Immobility and severe or profound intellectual disability reduce life expectancy.

- **Positive factors for independent living** include regular schooling, completion of secondary school, independent mobility and ability to travel beyond the house, good hand skills, living in a small community, and having spasticity as the motor dysfunction:
  - Intellectual disability, seizures, and wheelchair dependency are factors that reduce the likelihood of independent living.
- **Positive prognostic indicators for employment** include mild physical involvement, good family support, vocational training, and good employment contracts.

THERAPEUTIC MANAGEMENT

- The **Individuals with Disabilities Education Act (IDEA)** is a federal law that mandates *early intervention* for children who demonstrate developmental delay in their first 3 years of life (infants/toddlers under 3 years of age).
- Early intervention is a system of programs that work with the infant/toddler and family to optimize the development of the disabled child in society and enhance the family's capacity to meet their child's needs.
- PT/OT/speech therapy (ST) are part of the required developmental services that must be provided. There is no evidence that early intervention prevents disability or produces changes in brain organization. However, there is evidence that these strategies minimize secondary complications and support the families.

THERAPEUTIC EXERCISE METHODS

- Various exercise techniques are utilized during PT and OT and are described as follows: Please see Chapter 1, Stroke, for more details on rehabilitation methods for motor deficits.
- **Bobath Approach (most widely used):** Also known as neurodevelopmental treatment (NDT). The goal is to normalize tone, inhibit abnormal primitive reflex patterns, and facilitate automatic reactions with subsequent normal development.
- **Phelps:** Uses extensive bracing, withdrawing support as motion is performed with a minimum of tension, overflow, and substitution.
- **Deaver:** Uses extensive bracing, limiting all but two motions of an extremity.
- **Doman and Delacato:** Uses a series of set patterns repeated many times during the day, in an attempt to train cerebral dominance and normalization of function.
- **Rood:** Also known as Sensorimotor Approach. Emphasizes sensory and motor systems equally, activating muscles through sensory receptors.
- **Vojta-European method:** Activates postural development and equilibrium reactions to guide normal development.
- **Conductive education:** "Rehabilitation through learning"—system in which the child with motor dysfunction is expected to be a proactive participant and learner in overcoming the disability. The process is facilitated by a "conductor," who is trained to integrate the education and rehabilitation needs of the individual with goal-oriented group activities.
- **Patterning therapy:** A series of exercises and manipulations simulating prenatal and postnatal movements that are aimed at improving "neurologic organization" in children with brain damage and other congenital neurologic disabilities. Several published reviews of this theory have found it to be without merit and scientific support. Many clinicians consider this the same as Doman–Delacato.
- **Hippotherapy:** Therapy using characteristic movements of horses to provide motor and sensory input and has been linked to improvement of gross motor function.
- **SPIDER (Strengthening Program for Intensive Developmental Exercises and activities for Reaching health Capability):** An intensive therapy technique developed in 1993 to help individuals with neuromuscular disorders improve mobility, balance, and coordination through the use of elastic bungee cords that suspend the child in the exercise unit.

- **Aquatherapy:** Pool-based therapy with physiologic and mechanical effects that enable children with CP to perform activities they otherwise cannot perform on land.

SPASTICITY MANAGEMENT

- The mainstay of treatment is through a combination of a comprehensive PT/OT program (modalities, therapeutic exercise, ROM, casting, and splinting), medications, bracing, nerve/motor point injections, and surgery.
- Interventions such as nerve/motor point blocks and chemodenervation may be performed to reduce muscle tone in specific areas of the body. More aggressive measures include surgery.

Medications (Table 10–27)

- The management of spasticity in children with CP includes an increasingly wide assortment of medications. **Table 10–27** lists medications commonly used for the management of spasticity.

Bracing

- Supramalleolar orthosis (SMO):
  - Indicated to decrease pronation associated with hypotonia.
- Tone-reducing ankle–foot orthoses (TRAFOs) aid in gait by controlling the equinus or equinovarus deformity:
  - Designed to decrease abnormal reflexes
  - Come in various forms: Solid ankle, hinged or articulated, posterior leaf spring, floor reaction, or carbon fiber ankle–foot orthoses (AFOs).
  - *Full-length foot plate* extends past the toe to discourage toe flexion.
  - *Metatarsal support* discourages stimulation to a particularly reflexogenic area of the foot.
  - They are most effective during gait, but use during rest helps prevent contractures.
- Knee–ankle–foot orthoses (KAFOs):
  - KAFOs add direct control over knee flexion and extension, as well as varus and valgus foot deformities, but add bulk and weight.
- Hip–knee-ankle–foot orthoses (HKAFOs):
  - HKAFOs add direct control over hip position. KAFO and HKAFO braces do not significantly improve gait, but they do decrease deformity.

Nerve/Motor Point Blocks

- Nerve/motor point blocks are indicated for treatment of spasticity affecting specific muscle groups. They are commonly done to decrease hip adductor, hamstring, and gastrocnemius spasticity to correct the scissoring gait pattern and equinovarus foot deformity, as well as to avoid development of contractures.
- Phenol and alcohol are typical agents used for chemoneurolysis, both of which are neurolytic:
  - They cause a chemical neurectomy that is effective for 3 to 6 months.
  - Distal regeneration from the site of injection results in loss of effect after 4 to 6 months (less for motor points).
- An electrical stimulator and potentially US are used to identify the proper location for injection.
- Potential benefits: Prevention of deformity and improved function by facilitation of therapies, orthoses, and so on.
- *Disadvantages*: Temporary sensory dysesthesias (nerve block only), especially in the tibial and UE nerves; permanent weakness leading to deformity (e.g., tibial block leading to calcaneovalgus foot).
- After this procedure, an aggressive stretching and gait-training program is indicated.

TABLE 10–27 Medications for Management of Spasticity

MEDICATION	SITE OF ACTION	MODE OF ACTION	DOSE	SIDE EFFECTS	PRECAUTIONS	COMMENTS
Baclofen (oral)	GABA-B receptors in spinal cord	↓ release of excitatory neurotransmitters from afferent terminals	Start with 2.5–5.0 mg BID, increase by 2.5–5.0 mg every 3–5 days; max: 20 mg four times a day	Weakness, fatigue, confusion, constipation	May lower seizure threshold; abrupt withdrawal may precipitate seizures or hallucinations	Drug of choice for MS and SCI; oral form can be given when there is an acute baclofen pump failure
Baclofen (intrathecal)	GABA-B receptors in spinal cord	↓ release of excitatory neurotransmitters from afferent terminals	Test dose 50 mcg; pump dose 27–800 mcg/day	Weakness, fatigue, confusion, constipation, cardiorespiratory depression	May lower seizure threshold; abrupt withdrawal may precipitate seizures or hallucinations, cardiorespiratory arrest	Approved for MS, SCI, CP, TBI
Dantrolene	Intrafusal and extrafusal skeletal muscle fibers	↓ release of calcium from sarcoplasmic reticulum	Start with 0.5 mg/kg BID; increase by 0.5 mg/kg every 5–7 days; max: 12 mg/kg/day to 400 mg	Weakness, fatigue, drowsiness, diarrhea	Hepatotoxicity 2%; frequent liver function tests	Drug of choice for spasticity of cerebral origin
Benzodiazepines	Receptors of brainstem, reticular formation, spinal cord	↑ GABA-A binding, potentiating presynaptic inhibition	Start with 1–2 mg BID; increase by 1–2 mg every 2–3 days; max: 20 mg four times a day	Drowsiness, fatigue, impaired memory, and recall	Tolerance and dependence, CNS depression	Most helpful in incomplete SCI and immediate postoperative period in children with CP
Clonidine	Agonist in brain, brainstem, substantia gelatinosa of spinal cord	Inhibits short latency of motor neurons; augmentation of presynaptic inhibition	Start with 0.1 mg patch for 7 days	Bradycardia, hypotension, depression	Blood pressure and pulse monitoring	Effective in reducing spasms and resistance to stretch in MS and other central demyelinating conditions, SCI, stroke

TABLE 10–27 Medications for Management of Spasticity (*continued*)

MEDICATION	SITE OF ACTION	MODE OF ACTION	DOSE	SIDE EFFECTS	PRECAUTIONS	COMMENTS
Tizanidine	Adrenergic receptors both spinally and supraspinally	Prevents release of excitatory amino acids from presynaptic terminal of spinal interneurons; may facilitate glycine, an inhibitory neurotransmitter	Start with 2–4 mg at bedtime; increase 2 mg every 2–4 days; max: 36 mg	Dry mouth, sedation, dizziness	Orthostatic hypotension, hallucinations, elevated liver function tests	Dystonia, torticollis, blepharospasm, strabismus
Botulinum toxin	Presynaptically at the NMJ	Prevents release of ACh into NMJ by binding irreversibly to the presynaptic membranes of the ACh	1–12 units per kilogram depending on size of muscle; 50 units per site; no more frequent than every 3 months	Weakness, cramping, pain	Antibody formation (causes loss of effect), respiratory arrest (reported at higher doses) Targeted muscles No need for anesthesia	No sensory side effects No tolerance to repeat injections
Phenol	Peripheral nerve; motor endplate Denatures protein and causes nerve destruction with axonal degeneration; disrupts myoneural junctions	4%–6% aqueous solution; max: 20 mL	Pain, skin irritation, temporary sensory dysesthesias, peripheral neuropathy, permanent weakness	Anesthesia, cardiac arrhythmia, can cause scarring in muscle		Referred to as chemical neurolysis Can result in permanent dysesthesias

ACh, acetylcholine; BID, twice a day; CNS, central nervous system; CP, cerebral palsy; GABA, gamma-aminobutyric acid; MS, multiple sclerosis; NMJ, neuromuscular junction; SCI, spinal cord injury; TBI, traumatic brain injury.
Source: Modified from Stempien LM, Gaebler-Spira D. Rehabilitation of children and adults with cerebral palsy. In: Braddom RL, ed. *Physical Medicine and Rehabilitation*. W. B. Saunders; 1996:1113–1132, with permission.

EXPECTED EFFECTS OF SPECIFIC NERVE BLOCKS IN THE LOWER EXTREMITIES

- Obturator nerve blocks (anterior and posterior branches):
  - Reduce adductor tone.
  - Diminish scissor gait.
  - Promote passive abduction as a means of protecting hip joint integrity.
- Sciatic branch blocks to the medial hamstrings (semimembranosus and semitendinosus):
  - Lessen crouch gait and internal rotation deformities.
- Tibial blocks (and in recent past, tibial branch blocks to the heads of the gastrocnemius):
  - Diminish plantar flexion tone and allow better tolerance of AFOs.
- Femoral nerve blocks:
  - Diminish spastic recurvatum.

BOTULINUM TOXIN INJECTIONS

- Botulinum toxin affects the neuromuscular junction (NMJ) with essentially the same goal of reducing muscle tone. Specific muscles can be targeted with botulinum toxin injections.
- It has been used extensively to treat blepharospasm (focal dystonia of ocular muscles) and to treat torticollis.
- Botulinum toxin acts by irreversibly blocking presynaptic release of acetylcholine (ACh) into the NMJ:
  - Given as an IM injection, the onset of effect takes 24 to 72 hours and peaks at 2 weeks.
  - Days after exposure to the toxin, the axon fibrils begin to sprout and form junction plates on new areas of the muscle cell walls, rendering weakness reversible over a period of 3 months.
- Advantages over nerve blocks include:
  - Less technically demanding on clinicians
  - Does not cause dysesthesia
  - Decreased injection site pain and discomfort

Surgical Procedures

- Surgery may be indicated to improve function and appearance and to prevent or correct deformities.
- Surgical measures include neurosurgical procedures (selective dorsal rhizotomy [SDR] and intrathecal baclofen [ITB] pump placement) as well as orthopedic interventions (soft tissue releases, tendon lengthening, tendon transfers, joint fusions, or rotations/angulations).
- Note: Surgery to improve ambulation remains problematic.

NEUROSURGICAL PROCEDURES

- Neurosurgical procedures, such as SDR and ITB pump placement, have been employed in spasticity management.
- **Selective posterior rhizotomy**
  - SDR is designed to decrease the excitatory sensory input to the motor neuron, thereby decreasing spasticity.
  - The procedure consists of a laminectomy to expose the cauda equina.
  - The dorsal roots are electrically stimulated, and various criteria are used for determining which parts of the root contain more fibers involved with abnormal reflexes.
  - These rootlets are subsequently severed.
  - This technique allows for decreased tone without significant sensation loss.
  - Patient selection criteria include:
    - Lack of dystonia and/or athetosis
    - Preservation of functional strength independent of spasticity
    - Presence of selective motor control
    - Younger age (3–8 years old)
    - Lack of significant joint contractures and few previous orthopedic procedures
    - Cognitive preservation, motivation, and positive family supports are important.

  - Negative effects of SDR include:
    - Hypotonia (usually transitory immediately postop but occasionally lasts up to 6 months).
    - Weakness (unmasked by reduction of tone)
    - Sensory changes and bladder dysfunction (both usually of brief duration)
    - Hip dislocation (thought to be exacerbated by sparing of L1 root, leading to unbalanced hip flexor spasticity)
    - Lordosis possibly secondary to sparing of L1
    - After surgery, the children require an extensive PT/OT program to recover from postoperative weakness and to maximize functional gains.
  - Poor candidates for SDR include children with poor head and trunk control and children who use spasticity for functional purposes (i.e., extensor spasms to stand).
- **ITB pump placement:**
  - Also see Spasticity section in Chapter 12, Associated Topics in Physical Medicine and Rehabilitation.
  - A programmable pump delivers baclofen directly to the intrathecal space of the spinal cord (equivalent to chemically adjustable rhizotomy).
  - Allows for much lower doses of baclofen to be used, minimizing side effects.
  - Required to be refilled on a regular basis.

ORTHOPEDIC PROCEDURES

- Orthopedic interventions can be classified as either soft tissue or bony:
  - Soft tissue procedures are done at the muscle or tendon level and consist of either releases, lengthenings, or transfers.
  - Bony procedures consist of either joint fusions (ankle or spine) or osteotomies (e.g., [de] rotation osteotomy of the femur or tibia; angulation osteotomy of the femur).
- Rhizotomy and orthopedic surgery in combination are often required to gain the greatest improvement in gait.
- Gait lab analysis may help with determining the appropriate intervention.

AGING WITH CEREBRAL PALSY

- In general, health-related problems occur at about the same rate as in the regular population.
- One of the most common complaints is neck pain (second to osteoarthritic changes), occurring in 50% of spastic patients and 75% in the dyskinetic group.
- Scoliosis has a much higher incidence in nonambulatory individuals.
- Data suggest that individuals with CP are capable of near-normal reproduction.
- There is no correlation between degree of disability and level of sexual activity.

VOCATIONAL ASPECTS

- 🕮 Predictors of successful and unsuccessful employment include IQ level, ambulation level, speech, and degree of hand function.
- A European population-based study found that 33% of young adults participated in higher education (77% of controls) and 29% were competitively employed (82% controls).

Competitive (in the Work Force)/Able to Work

- IQ >80
- Ambulation with or without assistive device
- Speech hard to understand to normal
- Hand use normal to requiring assistance

Sheltered Employment

- IQ between 50 and 79
- Ambulation with or without assistive devices
- Speech hard to understand to normal
- Hand use normal to requiring assistance

Unemployable/Unable to Work

- IQ <50
- Nonambulatory and nonverbal
- Requires assistance using hand

REFLEX DEVELOPMENT (TABLE 10–28)

TABLE 10–28 Reflex Development

REFLEX	STIMULUS	RESPONSE	AGE OF SUPPRESSION	CLINICAL SIGNIFICANCE
Moro (startle)	Feeling of falling (sudden neck extension); unexpected loud noise	Shoulder abduction + shoulder, elbow, and finger extension, followed by arm flexion and adduction	4–6 months	Persists in CNS pathology, static encephalopathy
Rooting	Stroking lips or around mouth	Moving mouth, head toward stimulus in search of nipple	4 months	Diminished in CNS pathology, may persist in CNS pathology
Positive supporting	Light pressure or weight bearing on plantar surface	Legs extend for partial support of body weight	3–5 months replaced by volitional weight bearing with support	Obligatory or hyperactive abnormal at any age; early sign of lower extremity spasticity; may be associated with scissoring
Asymmetric tonic neck	Head turning to side	Extremities extend on face side, flex on occiput side ("fencer position")	6–7 months	Obligatory response abnormal at any age, persists in static encephalopathy
Symmetric tonic neck	Neck flexion	Arms flex, legs extend	6–7 months	Obligatory response abnormal at any age, persists in static encephalopathy
	Neck extension	Arms extend, legs flex		
Palmar grasp	Touch or pressure on palm or stretching finger flexors	Flexion of all fingers (hand fisting)	5–6 months	Diminished in CNS suppression, absent in LMN paralysis; persists/hyperactive in spasticity
Plantar grasp	Pressure on sole distal to metatarsal heads	Flexion of all toes	12–14 months when walking is achieved	Diminished in CNS suppression; absent in LMN paralysis; persists/hyperactive in spasticity
Automatic neonatal walking	On vertical support plantar contact and passive tilting of body forward side to side	Alternating automatic steps with support	3–4 months	Variable activity in normal infants; absent in LMN paralysis

(*continued*)

TABLE 10–28 Reflex Development (*continued*)

REFLEX	STIMULUS	RESPONSE	AGE OF SUPPRESSION	CLINICAL SIGNIFICANCE
Placement or placing	Tactile contact on dorsum of foot or hand	Extremity flexion to place hand or foot over an obstacle	Before end of first year	Absent in LMN paralysis or with lower extremity spasticity
Neck righting or derotative righting	Neck rotation in supine	Sequential body rotation from shoulder to pelvis toward direction of face	4 months replaced by volitional rolling	Nonsequential leg rolling suggests increased tone
Tonic labyrinthine	Head position in space, strongest at 45 degrees from horizontal supine or prone	Predominant extensor tone (while supine) Predominant flexor tone (while prone)	4–6 months	Hyperactive/obligatory abnormal at any age, persists in CNS damage/static encephalopathy

CNS, central nervous system; LMN, lower motor neuron.
Source: From Molnar Alexander MA, Matthews DJ, eds. *Pediatric Rehabilitation: Principles and Practice*. 4th ed. Demos Medical Publishing; 2010.

SPINA BIFIDA (MYELODYSPLASIA)

- Spina bifida is the most common type of neural tube defect (NTD) caused by congenital dysraphic malformations of the vertebral column and spinal cord.
- 220,000 infants are born with NTD annually worldwide, the majority occurring in developing countries due to malnutrition.
- Each year, approximately 1,400 babies born in the United States have spina bifida.
- It is the second most common childhood abnormality/disability disease after CP.
- It is also known as myelodysplasia, which is not to be confused with myelodysplastic syndrome (MDS), which refers to a group of hemopoietic disorders in the older population.

EPIDEMIOLOGY

- The highest incidence occurs in the British Isles, Ireland, Wales, and Scotland, and the lowest in Japan.
- In the United States, the rate of NTDs was about 0.6 per 1,000 births in 1989, with higher incidences in families of Irish, German, or Hispanic ancestry and lower among Asians and Pacific Islanders.

ETIOLOGY

- Both polycentric inheritance and environmental influences have been proposed. Increased familial incidence and recurrence rate and slightly greater number of affected females than males (1.2:1) point to a genetic etiology.
- Recurrence rate is 2.5% to 5% after the birth of one child with spina bifida and doubles after two affected children.
- Several environmental risk factors have been implicated:
  - Low socioeconomic class
  - Midspring conception
  - Maternal obesity

  - In utero exposure to anticonvulsant drugs (valproic acid, carbamazepine)
  - Maternal febrile illness
  - Nutrition (folic acid deficiency)
- Studies have demonstrated that folic acid (0.4 mg daily) taken periconceptually and during early pregnancy significantly reduces the occurrence and recurrence of NTDs.

PATHOGENESIS

- Theories about the pathogenesis of NTDs relate to embryonic development of the CNS.
- Neurulation of the anterior and posterior neuropores occurs during the third to fourth week after conception. The postneurulation phase takes place during weeks 4 to 7 postconception. Defects occurring during this phase are skin-covered lesions.
- Normal neural tube closure starts in the third week of gestation from the midcervical level and proceeds in both the cephalad and caudad directions.
- Defect of neural tube closure is thought to occur around day 26 and accounts for most lesions through midlumbar.
- The more distal caudal cell mass forms between days 26 and 30, eventually resulting in formation of the central canal in the embryonic tail.
- Caudal regression with rostral extension, resulting in fusion with the neural tube, results in formation of the spinal cord by day 53.
- Lesions of the lumbosacral levels occur before day 53.

PRENATAL DIAGNOSIS

- Elevated maternal alpha-fetoprotein (AFP) and acetylcholinesterase in the maternal serum and amniotic fluid as well as fetal US are methods of prenatal diagnosis. AFP is reliable in 80% of open NTDs in weeks 13 to 15.
- Amniocentesis done by weeks 16 to 18 is nearly 100% accurate for detecting elevated amniotic fluid AFP. Amniocentesis does not detect closed NTDs without leakage of fetal CSF.
- Fetal US between 16 and 24 weeks' gestation is reported to have >90% reliability.

TYPES OF SPINA BIFIDA (TABLE 10–29)

- The two major types are spina bifida occulta and spina bifida cystica.

Spina Bifida Occulta

- In spina bifida occulta, dysraphism affects primarily the vertebrae. The neural and meningeal elements are not herniated to the surface. A frequent sign in 50% of children is the presence of a pigmented nevus, angioma, hirsute patch, dimple, or dermal sinus on the overlying skin.
- Spina bifida occulta usually occurs in the lumbosacral or sacral segments. Unlike the cystic form, spina bifida occulta is not associated with Arnold–Chiari malformation.

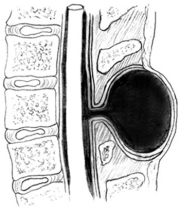
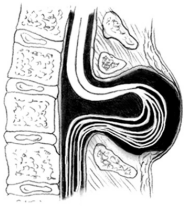
Spina Bifida Cystica

- Spina bifida cystica includes meningocele, myelomeningocele, myelocele, and other cystic lesions.
- In spina bifida cystica, contents of the spinal canal herniate through the posterior vertebral opening.
- The term "spina bifida aperta" refers to any NTD lesion in which the deformity is open to the environment.

CLINICAL SIGNS AND COURSE

- Clinical signs can be discerned by careful examination in the newborn nursery.
- Motor and sensory deficits vary according to the level and extent of the spinal cord involvement (**Table 10–30**).
- Motor paralysis is usually of the lower motor neuron (LMN) type.
- Sensory deficit is present in the dermatomes that would be innervated by the defective spinal segments and nerve roots.

TABLE 10–29 Types of Spina Bifida

	SPINA BIFIDA OCCULTA	SPINA BIFIDA CYSTICA		
		MENINGOCELE	MYELOMENINGOCELE	MYELOCELE
	Failure of fusion of the posterior elements of the vertebrae	The protruding sac contains meninges and spinal fluid 	The protruding sac contains meninges, spinal cord, and spinal fluid 	Cystic cavity is in front of the anterior wall of the spinal cord
Posterior elements of spine	Failure of fusion	Failure of fusion	Failure of fusion	Failure of fusion
Meninges herniate and form a cystic sac	No cystic sac formation	Cystic sac formation present	Cystic sac formation present	Cystic cavity is in front of the anterior wall of the spinal cord
Contents of cystic sac	No cystic sac formation	Spinal fluid Meninges	Spinal fluid Meninges Spinal cord	
Associated findings	50% of the children have: A pigmented nevus Angioma Hirsute patch Dimple or dermal sinus overlying skin	With or without intact skin at site of sac Incomplete skin coverage leads to leakage of CSF	Arnold–Chiari malformation Type II Complicated by hydrocephalus in over 90% of the cases—with or without intact skin at site of sac	
Clinical symptoms	No neurologic deficit Rarely associated with sacral lipoma and tethered cord; therefore, these children must be followed	In the absence of other underlying malformation, neurologic signs are normal, but children must be followed Meningocele occurs in <10% of cases of spina bifida cystica	Motor paralysis Sensory deficits Neurogenic bowel and bladder	
Spinal cord level involved	Lumbosacral or sacral region (most common level is L5 and S1 levels).	75% of these lesions affect the lumbar and lumbosacral segments. (The remainder are located in the thoracic or sacral area but only rarely at the cervical level.)	75% of these lesions affect the lumbar and lumbosacral segments. (The remainder are located in the thoracic or sacral area but only rarely at the cervical level.)	75% of these lesions affect the lumbar and lumbosacral segments. (The remainder are located in the thoracic or sacral area but only rarely at the cervical level.)
Population affected	Normal variant in approximately 5%–10% of the population.	Meningocele occurs in < 10% of cases of spina bifida cystica.	Myelomeningocele affects an overwhelming majority of the group with spina bifida cystica.	

CSF, cerebrospinal fluid.

SEGMENTAL INNERVATION

Musculoskeletal, Sensory, and Sphincter Dysfunction by Segmental Level (Table 10–30)

TABLE 10–30

	T6–T12	L1	L2	L3	L4	L5	S1	S2	S3	S4
Trunk	Abdominals Trunk flexion									
	Lower trunk extensors									
Hip		Iliopsoas Hip flexion								
		Hip adductors								
					Gluteus medius Hip abduction					
						Gluteus maximus Hip extension				
Knee			Quadriceps Knee extension							
					Hamstring-hip extension Knee flexion					
Ankle					Tibialis anterior Dorsiflexion, inversion					
						Peroneal eversion				
					Triceps surae Plantar flexion					
						Tibialis posterior Plantar flexion, inversion				
Foot					Toe extensors					
						Toe flexors				
						Foot intrinsics				
Perineum								Perineum sphincters		

	T6–T12	L1 L2 L3	L4 L5	S1	S2 S3 S4
Description	Complete leg paralysis Kyphosis Scoliosis Hip, knee flexion contractures Equinus foot Bowel and bladder dysfunction	Early hip dislocation Hip flexion and adduction contractures Scoliosis Lordosis Knee flexion contractures Equinus foot Bowel and bladder dysfunction	Late hip dislocation Scoliosis, lordosis Calcaneovarus or calcaneus foot Knee extension contractures Hip, knee flexion contractures Bowel and bladder dysfunction	Cavus foot Bowel and bladder dysfunction	Bowel and bladder dysfunction Cavus foot

Source: From Alexander MA, Matthews DJ, eds. *Pediatric Rehabilitation: Principles and Practice*. 5th ed. Demos Medical Publishing; 2015.

ASSOCIATED COMPLICATIONS OF SPINA BIFIDA

- **Arnold–Chiari malformation Type II (ACM II):**
  - Defined as downward displacement of the medulla and brainstem through the foramen magnum with associated kinking of the brainstem.
  - ACM II is present in almost all cases (80%–90%) of myelomeningocele and is complicated by hydrocephalus in over 90% of those cases.
- **Hydrocephalus:**
  - Usually present at birth and becomes symptomatic during the first week.
  - More than 80% of children require ventriculoperitoneal shunting with revision. Infection is the most frequent complication, followed by obstruction.
  - Spontaneous arrest of hydrocephalus occurs in 50% by 15 years, although "prophylactic" shunt removal is contraindicated. The presence of hydrocephalus correlates with spinal defect, thoracic greater than lumbar greater than sacral.
- **Vasomotor changes**—occur over involved area
- **Charcot joints**
- **Osteoporosis**
- **Malformations of forebrain and hindbrain**—decompression for symptomatic (ACM II) to minimize progression
- **Tethered cord**—abnormal attachment of spinal cord at the distal end:
  - Retethering occurs in 10% to 15%.
  - Typical signs and symptoms include increased weakness (55%), scoliosis (51%), pain (32%), orthopedic deformity (11%), and urologic dysfunction (6%).
- **Benign lumbosacral tumors:** Include lipoma and fibrolipoma
- **Diastematomyelia or sagittal cleavage (divisions) of the spinal cord:**
  - Usually associated with a bony spur
- **Syringomyelia:**
  - Fluid-filled central cavity (syrinx) in the spinal cord parenchyma lined with glial cells. Incidence is between 5% and 40%.
  - Usually occurs in the cervical spine
  - Presenting symptoms include deterioration of neurologic function, pain and temperature sensory deficits, loss of motor function especially of the UEs, increased spasticity and hyperreflexia, and pain.
  - MRI is best to evaluate if syringomyelia is suspected.
- **Scoliosis/kyphosis:**
  - Secondary to vertebral anomalies above the deficit and muscle weakness.
- 🕮 **Central respiratory dysfunction:**
  - Most frequent single cause of death in myelodysplasia. Symptoms include stridor, central apnea, and aspiration.
- **Dyscoordination:** Impaired fine hand coordination and ataxia
- **Impaired visual function:**
  - Strabismus, lateral rectus palsy, and nystagmus
- **Malformation of the urinary system:**
  - Renal hypoplasia, horseshoe kidney, solitary kidney, ureteral or lower tract anomalies
- **Skin breakdown:**
  - Contributory factors include excessive pressure, mental retardation, chronic soiling, and parental involvement.
  - Areas of breakdown:
    - Nonambulatory patients: Sacrum, ischial tuberosities, greater trochanters, and over gibbus deformity.
    - Ambulatory patients: The soles of the feet are the most common area.
- **Urinary dysfunction:**
  - >90% with myelomeningocele will have neurogenic bladder. Fewer than 10% have normal urinary control.

  - 🕮 T10–L2: Sympathetic adrenergic innervation
  - 🕮 S2–S4: Parasympathetic cholinergic innervation
  - 🕮 S2–S5: Somatic innervation through pudendal plexus
  - Disturbed bladder sensation interferes with perception of bladder filling.
  - Hypertonic (spastic) bladder most common in thoracic lesions.
  - Hypotonic (flaccid) bladder most common in sacral lesions.
  - Hydronephrosis is found in 7% to 30% of infants.
  - Urinary incontinence occurs in 95% of patients with spina bifida.
  - Detrusor sphincter dyssynergia is present in 50% of patients.
  - 🕮 Calcinosis is common with *Proteus* urinary tract infections.
- **Bowel dysfunction:**
  - There is autonomic innervation to the colon, rectum, and internal anal sphincter. Voluntary somatic motor and sensory nerve supply to the external sphincter S2–S4 occurs via the pudendal plexus.
  - Most children with spina bifida have fecal incontinence from poor rectal tone, absent cutaneous reflex response, and perianal sensory deficit.
  - 60% to 86% of children with spina bifida are dependent on their caregivers for bowel management.
  - In lesions above L2, intact spinal reflex arc can maintain sphincter tone despite absent rectal sensation.
  - Independence with toileting in myelomeningocele is the number one delayed self-care task, often delayed until 10 to 15 years of age.
  - 🕮 Presence of bulbocavernosus or anocavernosus reflex has been associated with a greater chance of bowel continence.
- **Obesity:**
  - Secondary to reduced daily energy expenditure and lower metabolic rate. Impedes ability to self-manage condition.
- **Precocious puberty:**
  - Secondary to premature activation of the hypothalamic–pituitary–gonadal axis from increased pressure on the hypothalamus.
  - Occurs in 10% to 20% of cases of myelomeningocele with hydrocephalus.
  - Breast and testicular enlargement occurs in those <8 to 9 years of age instead of the usual 11 to 11.6 years.
  - There is an associated short stature due to growth hormone deficiency. Treatment includes gonadotropin-releasing hormone analogs, although data on growth hormone are controversial.
- **Intellectual function:**
  - There is a three times higher incidence of low IQ scores in children with spina bifida.
  - 🕮 Intellectual function correlates inversely with the level of spinal cord dysfunction: The higher the lesion, the lower the IQ score.
  - Concentration and attention deficits are related to hydrocephalus.
  - Hydrocephalus alone does not exclude normal cognitive function. However, its complications, such as repeated CNS complications with meningitis, can lead to significant cognitive deficits.
  - Higher scores are achieved on verbal tasks than visual and visual motor activities.
  - Verbal skills are better than math, written, or visual perceptual abilities.
  - The term "cocktail party personality" represents children with good verbal ability that creates the impression of higher intellectual functioning than is found on formal testing.

TREATMENT AND MANAGEMENT

Neurosurgical Treatment

- Advances in minimally invasive prenatal repair of open spina bifida through fetoscopy.
- Neurosurgical repair of a cystic lesion is usually performed on the first day of life, with resultant lower mortality.

- Of these, 75% to 85% require shunting for hydrocephalus. The average revision rate of shunting is 30% to 50% for the first year, and 50% to 75% for the second year. Nearly all shunts are revised by 5 years of age.

Urologic Treatment

- Neurogenic bladders are seen in all patients except those with a very low sacral lesion and may be partial in low lumbar or high sacral lesions.
- Renal US is used to define anatomy in infancy (2 weeks old).
- GU malformations associated with spina bifida, although rare, include hypoplastic kidneys, horseshoe kidneys, renal agenesis, and ureteral duplications.
- Intermittent catheterization should begin when the residual volume is ≥20 mL.
- 🕮 Independent self-catheterization may be achieved at the age of 5 to 6 years, with boys learning intermittent catheterization more easily than girls secondary to anatomic differences. 15% to 20% have vesicourethral reflux at birth.
- Intravenous pyelogram (IVP) is recommended at about 2 years of age.
- Pharmacologic management includes anticholinergics that decrease detrusor contractions and enlarge bladder storage capacity, and alpha-adrenergic agents that increase outflow resistance.
- Surgical measures may include bladder augmentation using the ileum or colon, suprapubic vesicostomy, and artificial urinary sphincter.
- Culture and sensitivities are done initially at 6-week intervals but can be lengthened to 6-month intervals if symptom free.
- Long-term goal of bladder management is to prevent renal damage by preventing recurrent infections and reflux.

Orthopedic Management

- **Spine:**
  - Spinal deformities occur most commonly in thoracic lesions, with 80% to 100% of patients affected by the age of 14 to 15 years.
  - Kyphosis may be structural or paralytic.
  - Structural scoliosis may include vertebral anomalies, such as wedge or hemivertebrae, block-vertebral, and so on, and can occur alone or in combination. Scoliosis occurs secondary to loss of truncal support and is seen in 70% of patients above L2 and 40% below L4.
  - Treatment of spinal deformities:
    - Mild scoliosis: TLSO with regular follow-up
    - Rapidly progressing scoliosis: Check for tethered cord, which can cause rapid scoliosis and requires surgical correction.
    - Gibbous deformity: Kyphectomy
- **Hips:**
  - Hip dislocation and pelvic obliquity are common in paralytic scoliosis.
  - At hips, *bilateral dislocation* without restriction of joint mobility is best left alone.
  - *Unilateral dislocation* or asymmetric contractures may be treated as these can lead to pelvic obliquity, difficulty sitting, and decubiti.
- **Knees:**
  - Flexion or extension contractures of the knees are most common in thoracolumbar lesions. Knee flexion contractures ≤20 degrees are generally well tolerated in the ambulatory patient.
  - Serial casts can be tried early.
  - At the knees, soft tissue releases of flexion or extension contractures may increase mobility and allow sitting and/or bracing.
  - Osteotomies may be necessary late if shortening of the neurovascular bundle has occurred.
  - Tibial osteotomy for severe tibial torsion may be helpful.
- **Feet:**
  - Common deformities include equinus, equinovarus, calcaneal, cavus, and rocker-bottom feet.

- Rigid club foot is associated with thoracic or upper lumbar lesions.
- Posterior transfer of tibialis anterior is performed at >5 years of age for calcaneus foot deformity.
- Equinus deformity is often treated with Achilles tendon lengthening.
- Flexor tenodesis or transfer and plantar fascia fasciotomy are used to correct severe claw toe deformity and pes cavus.

Bowel Management

- Training with a timed bowel program may be initiated by 2 to 3 years of developmental age.
- Peristalsis and gastrocolic reflexes are usually intact, making postmeal evacuation most successful.
- Additional useful measures include stool softeners, bulk additives, suppositories, PRN enemas, digital stimulation, and manual removal.
- Surgical options include antegrade continence enema and colostomy.

Latex Allergy

- Latex sensitivity occurs in 59% of children with spina bifida and 55% in children who face multiple surgeries for other diagnoses.
- Predisposing factors are atopic disposition with known allergies, multiple surgical procedures, and previous frequent exposure to latex-containing gloves, nonsurgical equipment, or toys.
- Diagnostic tests include serum immunoglobulin E (IgE) antigen specific for rubber, skin pinprick test, and radioallergosorbent testing (RAST).
- Children should be carefully screened for any type of allergic reaction to latex.
- 🕮 There is a 500 times increased risk of anaphylaxis in the operating room for children with spina bifida.

MOTOR DEVELOPMENT IN SPINA BIFIDA

- During the first 6 months, motor development is close to normal with children attaining head control and hand play. From 6 to 12 months, delays become obvious, requiring adaptive equipment.

ROLLING

- Children with thoracic lesions usually roll by 18 months with compensatory strategies.
- Many with midlumbar deficits and all with L5 or sacral lesions get up on their hands and knees to crawl.

SITTING

- T12 lesions allow trunk control. Children with midlumbar lesions can usually sit with some delay and increased lordosis.
- If L4–L5 is spared, the child can sit normally.

PROGNOSTIC INDICATORS (TABLE 10–31)

- The level of neurologic impairment influences expectations for functional outcome and prognosis.

🕮 Ambulation Ability by Spinal Level

THORACIC LESIONS

- Children with thoracic lesions require assistive devices for passive standing, usually started at 12 to 18 months:
  - Assistive devices may include a parapodium, which allows sitting and standing, a swivel walker, HKAFOs with spinal extensions, and either a walker or Lofstrand crutches.
- Gait pattern is established from as low as drag to as high as swing through.

TABLE 10–31 Prognostication of Mobility in Myelomeningocele

MOTOR LEVEL SPINAL CORD SEGMENT	CRITICAL MOTOR FUNCTION PRESENT	MOBILITY: SCHOOL AGE	RANGE: ADULTS	ACTIVITY: ADOLESCENT
T12	Totally paralyzed lower limbs	Standing brace, wheelchair	Wheelchair	Wheelchair, no ambulation
L1–L2	Hip flexor muscles	Crutches, braces, wheelchair	Wheelchair, household ambulation	Wheelchair, nonfunctional ambulation
L3–L4	Quadriceps muscles	Crutches, braces, household ambulation, wheelchair	Crutches, household ambulation, wheelchair	50% wheelchair, household ambulation with crutches
L5	Medial hamstrings, anterior tibial muscles	Crutches, braces, community ambulation	Crutches, community ambulation	Community ambulation with crutches
S1	Lateral hamstring and peroneal muscles	Community ambulation	Community ambulation	Community ambulation 50% crutch or cane
S2–S3	Mild loss of intrinsic foot muscles possible	Normal	Normal	Limited endurance because of late foot deformities

Source: From Davis RD, Law C. Myelomeningocele and other spinal dysraphisms. In: Braddom RL, ed. *Braddom's Physical Medicine and Rehabilitation*. 4th ed. Elsevier, Sanders; 2011: chap 54, 1284, with permission.

LOWER THORACIC AND LUMBAR LESIONS

- Lower thoracic and lumbar lesions often require devices, such as a reciprocal gait orthosis (RGO), used after the age of 3 years. Tension is created by forward stepping, which generates an extension moment at the contralateral hip. Energy requirement is similar to wheelchair mobility.
- If L3 is spared, the child may be able to use an AFO.
- Children with low lumbar lesions pull to stand and cruise near the expected age. They walk around 2 years of age with Trendelenburg lurch and gastrocnemius limp. Functional community ambulation is realistic.
- The mental age of 2 to 3 years is a prerequisite to learn crutch walking. Low thoracic lesions and upper lumbar lesions may achieve crutch walking by 4 to 5 years.

LOW LUMBAR AND SACRAL LESIONS

- Bracing is usually not required, but patients may benefit from AFOs if plantar flexors are spastic or nonfunctional.

🕮 FUNCTIONAL COMMUNITY AMBULATION

- Thoracic lesions: 0% to 33%
- High lumbar: 31% achieve some degree of community ambulation.
- Low lumbar: 38% achieve functional community ambulation at <15 years of age:
  - 95% of functional community ambulators are 15 to 31 years of age.
- Sacral lesions: All are able to achieve functional community ambulation.

FACTORS/PREDICTORS FOR AMBULATION

- Sitting balance and motor level are early predictors of walking.
- Deformities of the spine and LEs and obesity are unfavorable factors for ambulation.
- Training in wheelchair use can begin during the second year. An electric wheelchair is recommended at school age for a child with adequate cognitive function and emotional maturity.

OUTCOMES

- Referral to preschool programs at age 3 years is legally mandated for children with disabilities. Most children with spina bifida complete high school, with 50% continuing to further education.
- Independent living is achieved by 30% to 60% of those with spina bifida in the United States.
- 🕮 The employment rate among those with spina bifida is 25% to 50%.
- Conception is possible in women; however, frequency of premature labor is increased.
- Male sexual function is present in those with L5 lesions and sacral deficits, with reproductive potential related to lower and less severe lesions.
- The incidence of spina bifida in offspring with one affected parent is 4%.
- Urologic causes are responsible for around 40% of deaths between 5 and 30 years, with survival during this period falling by 3% for every 5 years.

NEUROMUSCULAR DISEASES IN CHILDREN

Neuromuscular diseases are disorders that involve abnormalities, primarily of the LMN pathway (motor unit): Anterior horn cell, peripheral nerve, NMJ, or muscle **(Figure 10–20)**. Neuromuscular diseases may be acquired, but most commonly have a genetic etiology:

- Acquired neuromuscular diseases include acute inflammatory demyelinating polyneuropathy (AIDP) aka Guillain–Barré syndrome (GBS), poliomyelitis, botulism, myasthenia gravis (MG), polymyositis, and toxic neuropathies.
- The most common neuromuscular diseases are genetic. Examples include SMA, muscular dystrophies, hereditary motor sensory neuropathies (HMSN), and congenital MG.
- Molecular genetic advances have led to the discovery of specific genes for >100 neuromuscular disorders.
- Common symptoms of neuromuscular disease may include infantile floppiness or hypotonia, delay in motor milestones, feeding and respiratory difficulties, abnormal gait characteristics, frequent falls, difficulty with stairs or arising from the floor, and muscle cramps or stiffness.

CHARACTERISTICS ON PHYSICAL EXAMINATION

- 🕮 Pseudohypertrophy of muscles is seen in DMD and Becker's muscular dystrophy (BMD). Increased gastrocnemius calf circumference is caused by increase in fat and connective tissue, not actual muscle hypertrophy.
- Stork (or champagne bottle) leg appearance: Focal atrophy of distal LE muscles, particularly seen in HMSN.
- Muscle fasciculations: Seen in a variety of LMN disorders; especially common in SMA.
- Hepatosplenomegaly: Common in metabolic myopathy, such as acid maltase deficiency and Types 3 and 4 glycogenosis.
- Craniofacial changes and dental malocclusion: Common in congenital myotonic muscular dystrophy (MMD), congenital myopathies, congenital muscular dystrophy (CMD), and survival of motor neuron (SMN) Type II.

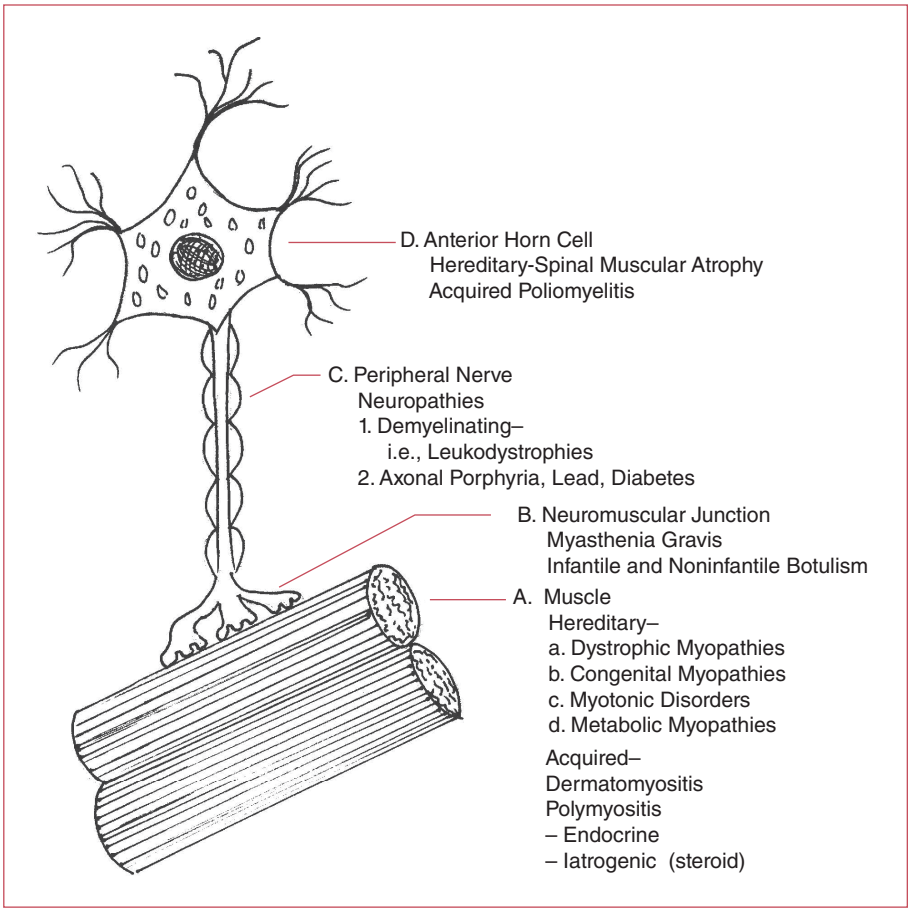


FIGURE 10–20
Anatomic breakdown of disorders of the lower motor neuron.

- 🕮 Gower's sign: Inability to rise from a seated position on the floor, normally due to proximal (pelvic girdle) muscle weakness:
  - Due to pelvic girdle weakness, patients are forced to rise off the floor by first rolling into a prone position, then assuming a 4-point stance (on hands and knees), followed by bridging the knees into extension while leaning the UEs forward (substituting for hip extension weakness by pushing off the knees with the UEs), and sequentially moving the UEs up the thigh until an upright stance with full hip extension is achieved.
  - Note: A positive Gower's sign is NOT specific to any one neuromuscular disease but indicative of proximal pelvic girdle muscle weakness.
- Myopathic gait: Hyperlordotic, waddling gait caused by weakness of back and hip extensors, which produces anterior pelvic tilt and a tendency for the trunk to be positioned anteriorly to the hip:
  - Patients compensate by maintaining lumbar hyperlordosis, which positions their center of gravity *posterior* to the hip and stabilizes the hip in extension.
  - Weakness of hip extensors also produces a tendency for knee instability and buckling:
    - Patients compensate by decreasing knee flexion and posturing the ankle into plantar flexion on initial contact during the stance phase of the gait cycle. This produces a knee extension moment at initial contact; plantar flexion of the ankle during mid to late stance helps to position the center of gravity anterior to maintain knee extension and knee stability.
  - Commonly seen in DMD and BMD
- Toe walking: This can be caused by paralytic muscle disease. The classic example is DMD. This progressive loss of muscle tissue is caused by fibrosis. Because the plantar flexor muscles are

approximately six times more powerful than the dorsiflexor muscles, the fibrosis leads to a fixed contracture in plantar flexion:
  - 🕮 Weakness in the quadriceps can also threaten knee stability. Compensation is achieved by walking on the forefoot, which maximizes the extension moment of the knee.
- Trendelenburg gait: Ipsilateral hip abductor (gluteus medius) weakness on the stance phase leg produces a pelvic drop on the swing phase (contralateral) side:
  - The patient compensates by bending the trunk laterally over the stance phase hip joint (compensated Trendelenburg gait).
- Steppage gait: Occurs secondary to distal weakness affecting ankle dorsiflexion. Steppage gait facilitates clearance of the plantar flexed ankle and foot with hyperflexion of the hip and knee joints.

SPECIFIC NEUROMUSCULAR DISEASES

- Neuromuscular diseases fall into four main categories:
  1. Myopathies
  2. NMJ disorders
  3. Peripheral nerve disorders
  4. Motor neuron disorders

1. Myopathies: Dystrophic, Congenital, Myotonic

Myopathies can be broken down into three different groups:

- **Dystrophic myopathies:**
  - DMD
  - BMD
  - CMD
  - Facioscapulohumeral (FSH) dystrophy
  - Emery–Dreifuss muscular dystrophy (EMD)
  - Limb girdle syndromes
- **Congenital myopathies:**
  - Mitochondrial myopathies
  - Central core myopathy
  - Minicore disease
  - Nemaline myopathy
  - Myotubular myopathy
  - Congenital fiber-type disproportion
  - Congenital Type 1 fiber predominance
  - Miscellaneous subcellular organelles
  - Nonspecific congenital myopathies
  - Minimal change myopathy
- **Myotonic myopathies:**
  - MMD
  - Myotonia congenita
  - Schwartz–Jampel syndrome
  - Congenital myotonic dystrophy

DYSTROPHIC MYOPATHIES

DUCHENNE MUSCULAR DYSTROPHY

A steadily progressive, X-linked muscular dystrophy that results from abnormality at the Xp^{21} gene loci and plasma membrane protein dystrophin deficiency. Chronic dystrophic myopathy is characterized by aggressive fibrotic replacement of the muscle and eventual failure of regeneration with muscle fiber death and fiber loss. Absent dystrophin or <3% of normal is diagnostic of DMD.

- *Incidence of DMD*: Around 1 per 3,500 to 5,500 male births. The majority of children are identified by 5 to 6 years old, most frequently with symptoms of abnormal gait, frequent falls, and difficulty climbing stairs.
- *Age of diagnosis*: Within the first 5 years of life for the first affected child
- *Presenting symptoms*:
  - Delay in walking
  - Abnormal gait
  - Frequent falling
  - Difficulty climbing steps
- *Cardinal clinical signs*:
  - Waddling gait, lordotic posture
  - Abnormal run
  - Difficulty rising from floor (Gower's sign)
  - Inability to hop
  - Proximal muscle weakness, legs > arms
  - Prominence of calves
- *Associated features*:
  - Cardiomyopathy (EKG abnormality)
  - Intellectual disability (variable)
  - Deformities—equinovarus, scoliosis after loss of ambulation, fixed flexion contractures after loss of ambulation
- *Course and prognosis*:
  - Progressive loss of function
  - Loss of ambulation, usually by 8 to 12 years
  - Prone to respiratory infections in later stages
  - Life expectancy: Late teens, early 20s
- *Investigations*:
  - Creatinine kinase (CK): Grossly elevated
  - EMG: Myopathic
  - US: Increased echo
- *Muscle biopsy*:
  - Progressive changes with time
  - Degeneration and regeneration, variation in fiber size, internal nuclei, proliferation of adipose and connective tissue
  - 🕮 Degenerating fibers are often observed in clusters, with necrotic fibers surrounded by macrophages and lymphocytes (Deconinck and Dan, 2007).
- *Genetics*:
  - X-linked recessive, gene location Xp^{21}
  - Counseling for carrier status of female relative based on CK and application of new advances in recombinant DNA technology
  - Prenatal diagnosis, chorionic villous biopsy
- *Management*:
  - Prevention of fixed deformities by passive stretching
  - Avoid immobilization with acute illnesses or injury
  - Promotion of ambulation with braces after loss of ability to stabilize hips and knees
  - Progression of scoliosis will destabilize sitting posture, and the provision of spinal supports and appropriate seating will stabilize trunk, facilitate arm use, and protect skin. Spine surgery should be considered early when the curvature measures 20 degrees or more.
  - Maintain strength and prolong ambulation with glucocorticoids including prednisone or deflazacort
  - Operative treatment of progressive scoliosis
  - Glucocorticoids are used in the treatment of DMD, targeting symptoms and disease progression by reducing inflammation and progressive degeneration of skeletal and cardiac muscles.

Prednisone may slow weakness progression and prolong ambulation by 2 to 3 years. It may also improve pulmonary and cardiac function, especially if used early by age 2 to 4.
  - Prednisone/prednisolone and deflazacort are the steroids most used in the treatment of DMD.
  - Deflazacort, a derivative of prednisolone, was approved for DMD patients aged 5 and older. Though comparable, it may demonstrate greater delay in loss of ambulation, and cardiac, pulmonary, and motor decompensation.
  - Significant side effects may be associated with long-term use and include weight gain, adrenal insufficiency, cushingoid appearance, changes in personality, decreased bone density, stunted growth, and tendency for bone fracture. Short-term intermittent use is suggested.
  - Other novel dissociative steroids are being explored in clinical trials which may provide increased benefit of decreasing steroid-associated adverse effects.
  - Novel therapies for dystrophin restoration (useful in 13% of DMD patients with nonsense mutation): Ataluren, Drisapersen, Eteplirsen
  - Gene therapy: Elevidys (delandistrogene moxeparvovec-rokl), for ambulating patients aged 4 to 5 with mutation DMD gene
- 🕮 *Order of muscle group involvement:* Earliest weakness is seen in neck flexors during the preschool years. Although early on the weakness is generalized, it is predominantly proximal. Pelvic girdle weakness precedes shoulder weakness by several years. Ankle dorsiflexion is weaker than ankle plantar flexion. Ankle evertors are weaker than inverters. Knee extensors are weaker than flexors. Hip extensors are weaker than flexors. Hip abductors are weaker than adductors.
- >40% to 50% loss of muscle power occurs by age 6 years.
- The average age to wheelchair dependency is approximately 10 years with a range of 7 to 13 years. One study showed that all DMD subjects who took ≥9 seconds to ambulate 30 feet lost ambulation within 1 year.
- Contractures are common in children >13 years of age and mostly affect ankle plantar flexors, knee flexors, hip flexors, iliotibial band, elbow flexors, and wrist flexors.
- *Scoliosis*: Prevalence varies from 33% to 100% and is related to age. 50% acquire scoliosis between ages 12 and 15 years. Scoliosis usually develops after 3 to 4 years in the wheelchair though no cause–effect relationship has been established.
- *Pulmonary*: Forced vital capacity (FVC) volumes increase during the first decade of life and plateau during the early part of the second decade. There is a linear decline of FVC between age 10 and 20 years old. Peak obtained FVC usually occurs in the early part of the second decade and is an important prognostic indicator of severity of spinal deformity and severity of restrictive pulmonary compromise secondary to muscular weakness. FVC <40% is a contraindication to surgical spinal arthrodesis because of increased perioperative morbidity.
- *Cardiac*: Dystrophin protein is present in myocardium and cardiac Purkinje fibers; therefore, cardiac symptoms are part of the clinical picture. Nearly all patients >13 years old demonstrate EKG abnormalities. These may include Q-waves in the lateral leads, elevated ST-segments, poor R-wave progression, increased R/S ratio, and resting tachycardia and conduction defects. Cardiomyopathy usually is first noticed at age >10 years old and is apparent in nearly all patients >18 years old. Cardiomyopathy may result in GI-like symptoms in young patients; therefore, careful diagnostic monitoring is suggested in this population. Cardiomyopathy and systolic dysfunction are poor prognostic indicators.

BECKER'S MUSCULAR DYSTROPHY

This X-linked muscular dystrophy has a similar clinical pattern and gene locus to Duchenne type but is milder with slower progression.

- Unlike DMD, dystrophin is present at 20% to 80% normal levels or normal quantity in BMD. The dystrophin molecular weight is abnormal (usually reduced or increased) and results in anomalous function.

- Becker's has a later onset and a slower rate of progression compared with DMD.
- Prevalence of BMD is 12 to 27 per million, with a lower incidence than DMD.
- BMD patients are able to walk into the late teenage years. In DMD, ambulation is arrested earlier.
- BMD and DMD have similar distributions of weakness.
- *Age of onset*:
  - Variable: Usually after 5 years of age and into adolescence or adult life.
- *Presenting symptoms*:
  - Difficulty with running or climbing steps
  - Cramps on exercise
- *Cardinal clinical signs*:
  - Mild functional disability
  - Proximal muscle weakness
  - Prominence of calves
  - Waddling gait, lordosis
- *Associated features*:
  - Cardiac involvement (mild, variable EKG changes). Around 75% of Becker's patients have EKG abnormalities.
- *Course and prognosis*:
  - Slowly progressive, variable course compared with DMD
  - Some cases are practically static
  - Ambulation beyond 16 years
  - Life expectancy dependent on degree of progression and late respiratory deficit
- *Investigations*:
  - CK: Grossly elevated (similar levels to DMD)
  - EMG: Myopathic
  - US: Increased echo (variable)
- *Muscle biopsy*:
  - Variable dystrophic changes; degeneration and regeneration
  - Variable loss of fibers and proliferation of adipose or connective tissue; foci of atrophic fibers resembling denervation
- *Genetics*:
  - X-linked recessive, same locus (Xp^{21}) at DMD
  - Genetic counseling of heterozygote female carriers on basis of CK, as well as recombinant DNA technology
- *Management*:
  - Promotion of activity
  - Prevention of fixed deformity (e.g., equinus) by passive stretching
  - Braces for promotion of ambulation if loss of ability to walk in late stages
  - Prevention and management of scoliosis if chair-bound (Dubowitz, 1978)

CONGENITAL MUSCULAR DYSTROPHY

A heterogeneous group of cases presenting with clinical weakness or deformities in early infancy and having variable dystrophic changes in the muscle.

- Infants present with hypotonia, muscle weakness at birth or within the first few months of life, congenital contractures, and a dystrophic pattern on muscle biopsy.
- Children exhibit early contractures, equinovarus deformities, knee flexion contractures, hip flexion contractures, and tightness of wrist flexors and long finger flexors.
- *Age of onset*:
  - At birth or in infancy or early childhood
- *Presenting symptoms*:
  - Hypotonia and weakness

  - Fixed deformities (arthrogryposis)
  - Variable sucking, swallowing, and respiratory difficulty
  - Delayed motor milestones in later onset cases
- *Cardinal clinical signs*:
  - General hypotonia and weakness
  - Fixed deformities in relation to intrauterine posture
  - Variable weakness or contractures in later presenting cases
- *Associated features*:
  - Intellectual disability (especially in Japan—Fukuyama type)
  - Dislocation of hips
  - Secondary deformities, for example, scoliosis
  - Hydrocephalus and fundal changes (Santavuori type)
- *Course and prognosis*:
  - Variable—many cases relatively static
  - May show functional improvement with time
  - May be fatal from respiratory deficit and risk of superimposed infection
- *Investigations*:
  - CK: Variable from moderate elevation to normal levels
  - US: Marked increase in muscle echo
  - EMG: Myopathic pattern
- *Muscle biopsy*:
  - Variable–some show extensive dystrophic changes with marked replacement of muscle by adipose tissue and variable connective tissue proliferation.
  - Others can show mildly myopathic/dystrophic changes.
- *Genetics*:
  - Autosomal recessive, gene 9q31–33; 6q.
  - Some cases are probably sporadic.
  - Some may be sequel to viral or other inflammatory process.
- *Management*:
  - Active physiotherapy to encourage mobility
  - Passive stretching of "fixed" deformities (especially two joint muscles)
  - Surgical correction of residual deformities at appropriate stage (e.g., equinovarus correction when able to stand)
  - Avoid immobilization that promotes fixed deformity.
  - Supportive treatment for respiratory problems

FACIOSCAPULOHUMERAL MUSCULAR DYSTROPHY

Autosomal dominant dystrophy disorder primarily affecting facial and shoulder girdle muscles.

- Initial weakness affects facial muscles, especially the orbicularis oculi, zygomaticus, and orbicularis oris. The masseter, temporalis, extraocular, and pharyngeal muscles characteristically are spared.
- Sensorineural hearing deficit and impaired hearing function are more common than expected in FSH.
- Posterior and lateral scapular winging, high riding scapula, and hyperlordosis are also seen. Hyperlordosis occurs in 20% of patients with FSH. FSH patients with scoliosis have mild, nonprogressive curves.
- Mild restrictive lung disease occurs in nearly 50% of patients with FSH, with expiratory muscles more affected than inspiratory muscles.
- Cardiac complications in FSH are rare and patients generally have normal longevity.
- There is usually no cognitive deficit.
- *Age of onset*:
  - Variable, ranging from early childhood to adult life

- *Presenting symptoms*:
  - Some cases have trunk and pelvic girdle weakness and difficulty with locomotion.
  - Disability relating to shoulder or facial muscles
- *Cardinal clinical signs*:
  - Facial weakness—patient cannot whistle
  - Scapular winging
  - Shoulder girdle weakness
  - "Terracing" of shoulders on abduction
  - Lordosis and pelvic girdle weakness in some families
- *Associated features*:
  - Deafness (variable)
  - Fundal changes (variable)
- *Course and prognosis*: Very variable:
  - Some may be mild and very slowly progressive, with a normal life span.
  - Some have more marked progression of lower limb weakness and may lose ambulation in adult life.
  - Variable degree of facial muscle weakness
  - Variable respiratory deficit in later stages
- *Investigations*:
  - CK: Normal or slightly elevated
  - EMG: Normal or myopathic
  - US: Variable
- *Muscle biopsy*:
  - Variable pathologic change from focal atrophic fibers only to overtly dystrophic picture with variability in fiber size, splitting of fibers, internal nuclei, and proliferation of connective and adipose tissue.
  - Some cases have marked inflammatory response.
- *Genetics*:
  - Autosomal dominant inheritance, gene locus 4q35
  - Marked clinical variability within families. Subclinical cases may occur.
  - Genetic counseling needs careful clinical assessment of all family members to exclude subclinical members.
- *Management*:
  - Promotion of activity
  - Some cases benefit from surgical fixation of the scapulae to facilitate abduction of the arms (Dubowitz, 1978).

EMERY–DREIFUSS MUSCULAR DYSTROPHY

This X-linked muscular dystrophy is clinically distinct from Duchenne and Becker's types.

- Emerin is the muscle protein deficient in EMD.
- EMD usually presents in adolescence or early adulthood with atrophy in the upper arms and legs due to focal wasting of the calf muscles and biceps.
- The clinical hallmark of EMD is early presence of contractures of the elbow flexors with limitation of full elbow extension. Heel cord tightness with ankle dorsiflexion weakness and toe walking may also be present.
- Tightness of cervical and lumbar spinal extensor muscles, resulting in limitation of neck and trunk flexion, may occur.
- *Age of onset*:
  - Late childhood, adolescence, or adult life
- *Presenting symptoms*:
  - Difficulty with walking/running
  - Rigidity of neck or spine
  - Cardiac arrhythmia

- *Cardinal clinical signs*:
  - Early presence of contractures of the elbow flexors with limitation of full elbow extension
  - Fixed deformities: Equinus of feet, flexion deformity of elbows, rigidity of spine with limited neck and trunk flexion
  - Mild weakness
  - Focal wasting of muscles, especially upper arm (biceps and triceps) and lower leg (gastrocnemii, anterior group)
- *Associated features*:
  - Cardiac arrhythmia, may not be obvious clinically or on routine EKG, needs 24-hour Holter monitoring
  - Nocturnal hypoventilation, respiratory problems
- *Course and prognosis*:
  - Muscle weakness and functional disability, very slowly progressive
  - Cardiac involvement may be life-threatening in early adult life.
- *Investigations*:
  - CK: Slight to moderate elevation
  - EMG: Myopathic
  - US: Focal involvement with increase in echo
- *Muscle biopsy*:
  - Mild dystrophic changes with variability in fiber size, internal nuclei, proliferation of connective tissue, degeneration, or regeneration
  - Foci of atrophic fibers resembling denervation
- *Genetics*:
  - X-linked recessive, gene locus Xq^{28}
  - Gene not near Duchene and Becker's dystrophy genes
  - Counseling of heterozygote females on basis of CK elevation, minor changes on muscle biopsy, and possible DNA polymorphisms in future
- *Management*:
  - Promotion of ambulation
  - Prevention of deformities or their progression
  - Correction of fixed deformities of ankles if ambulation is affected
  - Close monitoring of cardiac status—may need cardiac pacemaker
  - Assessment of respiratory function (Dubowitz, 1978)

LIMB GIRDLE SYNDROMES

Autosomal recessive muscular dystrophy of variable severity that resembles BMD and DMD. These are myopathies characterized by predominantly proximal weakness of shoulder and pelvic girdle muscles.

- *Age of onset*:
  - Wide, from early childhood to adolescence and adult life
- *Presenting symptoms*:
  - Difficulty with gait, running, or climbing steps
  - Cramps on exercise
- *Cardinal clinical signs*:
  - Abnormal gait, lordotic posture
  - Functional disability with hopping and rising from floor
  - Variable muscle weakness
  - Deformities after loss of ambulation, as in DMD
  - Prominence of calves in some
- *Associated features*: No consistent ones
- *Course and prognosis*: Very variable:
  - Progression is usually slow, but some cases can be more severe and even more rapid than Duchenne type.

- *Investigations*:
  - CK: Elevation variable—mild to moderate, sometimes gross
  - EMG: Myopathic findings
  - US: Variable increase in echo; may show differential muscle involvement
  - Chromosome analysis in female cases to exclude translocation involving Xp21 site
- *Muscle biopsy*:
  - Dystrophic changes; variable
  - May be marked variability in fiber size and splitting of fibers
  - Degeneration and regeneration
  - Proliferation of adipose and connective tissue
- *Genetics*:
  - Autosomal recessive, gene 15q
- *Management*:
  - Promotion of ambulation
  - Prevention and treatment of deformities (Dubowitz, 1978)

CONGENITAL MYOPATHIES

This group of disorders usually presents with infantile hypotonia as a result of primary myopathies that are caused by genetic defects of the muscle fibers. There is no structural abnormality of the CNS or peripheral nervous system.

CENTRAL CORE MYOPATHY

- Autosomal dominant disorder with gene locus at 19q13.1
- High incidence of malignant hyperthermia with inhalation of anesthetic agents
- Predominance of Type I muscle fibers (high-oxidative, low-glycolytic) and relative paucity of Type II fibers, resulting in relative deficiency of glycolytic enzymes
- Patients demonstrate mild relatively nonprogressive weakness.

MINICORE DISEASE

- Autosomal recessive inheritance with predominance of Type I fiber involvement.
- Present with hypotonia, delayed motor development, nonprogressive symmetric weakness of trunk and proximal limbs, and diaphragmatic weakness with risk of nocturnal hypoventilation.

NEMALINE MYOPATHY (ROD-BODY MYOPATHY)

- Most typically occurs as autosomal recessive. An autosomal dominant gene has also been localized to chromosome 1q21–q23.
- Most cases present with a mild nonprogressive myopathy with hypotonia and proximal weakness.

MYOTUBULAR MYOPATHY

- Can be autosomal dominant/recessive or X-linked recessive (Xq^{28}) inheritance.
- Most common type of centronuclear myopathy (muscle fiber nuclei found at the center of the cell, as opposed to the periphery where they are usually found).
- Autosomal inherited types present with hypotonia, delay in motor milestones, generalized weakness of proximal and distal musculature, and ptosis with weakness of external ocular muscles and axial muscles.
- X-linked inherited type presents with neonatal onset of severe generalized hypotonia, severe muscle weakness, dysphagia, and respiratory insufficiency, often requiring ventilator support at birth.

MYOTONIC MYOPATHIES

- Myotonic myopathies are muscular dystrophies that are characterized by delayed relaxation (sustained contraction) of muscles after contraction (myotonia). These include diseases such as MMD, myotonia congenita, Schwartz–Jampel syndrome, and congenital myotonic dystrophy.

MYOTONIC MUSCULAR DYSTROPHY: STEINERT'S DISEASE OR DYSTROPHIA MYOTONICA

Autosomal dominant muscular dystrophy with an incidence of 1 per 8,000. Syndrome comprised of myotonia, muscle weakness/wasting, cataracts, premature balding, cardiomyopathy with conduction deficits, gonadal atrophy, and variable intellectual deficit and dementia. This adult type is the most common, but it may start in childhood. In addition, there is a distinct congenital type (most severe form):

- Myotonia can be seen with grip myotonia of the hand and percussion myotonia.
- It affects skeletal muscle, smooth muscle, myocardium, brain, and ocular structures. Associated findings include frontal baldness and gonadal atrophy, cataracts, and cardiac dysrhythmia.
- Characteristic facial features include long, thin face with temporal and masseter muscle wasting.
- MMD characteristically exhibits greater distal than proximal weakness, with initial weakness often in the ankle dorsiflexors, evertors, inverters, and hand muscles.
- Cardiac involvement is common, with EKG abnormalities in 70% to 75% of patients, with sudden death in <5% of patients.
- IQ is often significantly reduced.
- *Age of onset*:
  - Usually adolescence/adult. May be present in at-risk families at an early age.
- *Presenting symptoms*:
  - Weakness
  - Stiffness
- *Cardinal clinical signs*:
  - Voluntary myotonia with sustained grip
  - Percussion myotonia of tongue or peripheral muscles
  - Facial weakness, inability to bury eyelashes
  - Ptosis, frontal balding, hatchet facies
- *Associated features*:
  - Cataracts
  - Delayed intellectual development
- *Course and prognosis*:
  - Affected children identified in at-risk families are often symptom free. They may later develop the full adult syndrome, but severity is extremely variable, even within a family.
  - Prognosis depends on associated cardiomyopathy and respiratory problems.
- *Investigations*:
  - EMG: Myotonia plus myopathy
  - EKG: Conduction defects, arrhythmia
  - Ultrasonography: Increased muscle echogenicity
- *Muscle biopsy*:
  - In full-blown adult type, marked dystrophic changes plus internal nuclei and sarcoplasmic masses
- *Genetics*:
  - Autosomal dominant with marked clinical heterogeneity, gene 19q13
- *Management*:
  - Supportive treatment of dystrophy
  - Encourage activity
  - Myotonia not usually a problem (Dubowitz, 1978)

MYOTONIA CONGENITA: THOMSEN'S DISEASE (LITTLE HERCULES)

Autosomal dominant disorder inherited as a dominant or recessive trait in which myotonia is the only or predominant feature. Symptoms usually present after birth:

- Symptoms are exacerbated by prolonged rest or inactivity and cold.
- Muscle hypertrophy is commonly seen.

- *Age of onset*:
  - Any time from birth through infancy and childhood
- *Presenting symptoms*:
  - Stiffness after period of rest or sustained activity or posture
  - Stiffness and immobility on walking
  - Stiffness in cold weather
  - Difficulty releasing grip or object
  - Sustained eye closure in crying infant
- *Cardinal clinical signs*:
  - Myotonia following hand closure, eye closure, ocular deviation, or other sustained movements
  - Percussion myotonia of tongue, thenar eminence, or other muscles
  - Normal muscle power and function (usually)
  - Muscle hypertrophy
- *Associated features*:
  - Myotonia following fright or sudden muscle tension
  - Persistent weakness in some cases
  - Overlap with hyperkalemic periodic paralysis in some families
- *Course and prognosis*:
  - Usually good. Patient can often control myotonia by activity.
  - Tendency to improve with time
- *Investigations*:
  - EMG: Characteristic myotonic discharges (sound like dive bomber), on insertion of the needle
- *Muscle biopsy*:
  - Essentially normal
  - Fiber hypertrophy may be present.
- *Genetics*:
  - There is a recessive disorder of myotonia congenita, which is probably more common than the dominant disorder (gene 7q35). Question as to whether paramyotonia congenita is a separate genetic entity or a variant of myotonia congenita.
- *Management*:
  - Most patients manage without medication. When symptoms become disabling, therapeutic benefit may be obtained from quinine, procainamide, phenytoin, corticosteroids, or tocainide (Dubowitz, 1978).

SCHWARTZ–JAMPEL SYNDROME

- Autosomal recessive disease with symptoms of hypotonia, dwarfism, diffuse bone disease, narrow palpebral fissures and blepharospasm, micrognathia, and flattened facies.
- Symptoms are nonprogressive.

CONGENITAL MYOTONIC DYSTROPHY

- A type of myotonic dystrophy occurring in infants who are almost always born to mothers affected with myotonic dystrophy. It is characterized by severe hypotonia at birth, facial weakness, arthrogryposis, and variable breathing and swallowing difficulties.
- *Age of onset*:
  - In utero. Poor movements may be noted in second trimester. Always manifest at birth.
- *Presenting symptoms*:
  - Floppiness
  - Poor sucking and swallowing
  - Respiratory insufficiency
- *Cardinal clinical signs*:
  - Marked hypotonia

  - Variable weaknesses; may have antigravity power in limbs
  - Facial weakness
- *Associated features*:
  - Talipes equinovarus
  - Other deformities, depending on in utero posture
  - Hydramnios, premature bladder
  - Thin ribs (x-ray)
  - Ventricular dilation, with or without periventricular hemorrhage
  - Delayed intellectual development
- *Course and prognosis*:
  - Respiratory insufficiency may be life-threatening, particularly if compounded by marked prematurity.
  - If infant survives newborn period, swallowing difficulty resolves.
  - Hypotonia gradually resolves.
  - Marked delay in motor milestones
  - Variable delay in intellectual milestones
  - Speech difficulties
- *Investigations*:
  - Examine mother for facial weakness and myotonia
- *Muscle biopsy*:
  - May be histologically normal
  - Delayed maturation on electron microscopy and immunocytochemistry
- *Genetics*:
  - Dominant inheritance; mother always affected in congenital syndrome
  - Only infants with gene get hypotonic syndrome.
  - Further affected children likely to have congenital syndrome
  - Prenatal diagnosis with DNA probes possible in some families
- *Management*:
  - Respiratory support and tube feeding if appropriate to severity in infant
  - Passive stretching for equinovarus and other fixed deformities
  - Delay orthopedic intervention for talipes until child is standing
  - Speech therapist to advise on feeding difficulties and speech problems
  - Schooling appropriate to intellectual development (Dubowitz, 1978)

MITOCHONDRIAL MYOPATHIES (METABOLIC)

- Mitochondria are protein structures within a cell that produce cellular energy in the form of adesonine triphosphate (ATP). Mitochondrial myopathies are a group of metabolic neuromuscular diseases caused by genetic defects that result in defective mitochondria.
- Mitochondrial genetics are complex, as both nuclear and mitochondrial DNA gene mutations can affect the mitochondria:
  - Inheritance patterns are typically either autosomal recessive or maternal pattern. However, because a cell can contain both defective and normal mitochondria, disease severity can be variable even in the same family.
- Common mitochondrial myopathies:
  - Kearns–Sayre syndrome (KSS)
  - Leigh syndrome and maternally inherited Leigh syndrome (MILS)
  - Mitochondrial DNA depletion (MDS) syndrome
  - Mitochondrial encephalomyopathy, lactic acidosis, and stroke-like episodes (MELAS)
  - Mitochondrial neurogastrointestinal encephalomyopathy (MNGIE)
  - Myoclonus epilepsy with ragged red fibers (MERRF)
  - Neuropathy, ataxia, and retinitis pigmentosa (NARP)
  - Pearson syndrome
  - Progressive external ophthalmoplegia (PEO)

OTHER CONGENITAL MYOPATHIES

- Congenital fiber type disproportion—recessive
- Congenital Type 1 fiber predominance—recessive
- Miscellaneous subcellular organelles
- Nonspecific congenital myopathies, minimal-change myopathy (Dubowitz, 1978)

2. Neuromuscular Junction Disorders

TRANSIENT NEONATAL MYASTHENIA

- A transient disorder in a potentially normal infant of a mother with MG.
- Occurs in 10% to 15% of infants born to myasthenic mothers and is secondary to placental transfer of circulating ACh receptor (AChR) antibodies from mother to fetus with symptoms appearing within the first few hours of birth.
- Clinical signs include difficulty feeding, generalized weakness and hypotonia, respiratory difficulties, fetal cry, facial weakness, and ptosis.
- Usually self-limiting, with resolution in approximately 2 to 3 weeks.

🕮 CONGENITAL OR INFANTILE MYASTHENIA

- This occurs in the infants of nonmyasthenia mothers and may have an autosomal recessive inheritance. AChR antibodies are usually absent. There may be a number of different mechanisms involved.
- Pyridostigmine and 3,4-diaminopyridine approved for increasing ACh in congenital myasthenic syndromes (CMS).

JUVENILE MYASTHENIA

- This is similar to the adult autoimmune type and has a high titer of AChR antibodies. It particularly affects adolescent girls and is often severe and labile.
- *Clinical features*:
  - Ptosis and ophthalmoplegia are commonly present, together with weakness of other muscle groups including the face, jaw, swallowing, speech, respiration, and also neck, trunk, and limb muscles.
- *Investigations*:
  - 🕮 Fatigability of muscle (surface electrodes) after repetitive stimulation of the peripheral nerve (surface electrodes) at 4 or 10 Hz
  - Response to intravenous edrophonium (Tensilon®)
  - Single-fiber EMG, miniature endplate potentials (difficult in children)
- *Management*:
  - Emergency intensive treatment for bulbar and respiratory paralysis
  - Neonatal MG: Prostigmine or pyridostigmine; self-limiting
  - Congenital/infantile MG: Pyridostigmine
  - Juvenile (with Ab to AChR): Comprehensive regimen of prostigmine, plasma exchange, thymectomy, alternate day steroid therapy, and azathioprine, carefully tailored to achieve and maintain full remission (Dubowitz, 1978)

AUTOIMMUNE MYASTHENIA GRAVIS

- Proximal muscles are more affected than distal, and upper limbs greater than lower limbs.
- MG typically presents with ophthalmoparesis, ptosis, facial weakness, dysphagia, speech problems, neck, trunk, and proximal limb weakness, as well as often respiratory difficulties.
- Relapses and remissions are common.
- Diagnosis is confirmed by response to an anticholinesterase drug such as edrophonium (Tensilon).
- Repetitive nerve stimulation studies show a characteristic decrement in compound motor action potential (CMAP) amplitudes at slow stimulation rates (2–5 Hz) over a train of four to five stimuli. Decrements >10% are noted.

- Anticholinesterase receptors are detected in around 85% to 90% of patients with generalized MG and around 50% with ocular myasthenia.

INFANTILE BOTULISM

- This usually presents between 10 days and 6 months, with acute onset of hypotonia, dysphagia, constipation, weak cry, and respiratory insufficiency.
- Examination reveals weakness, ptosis, ophthalmoplegia with pupillary dilatation, decreased gag reflex, and preservation of deep tendon reflexes (DTRs).
- Diagnosis is by EMG or measurement of *Clostridium botulinum* toxin in the stool.

NONINFANTILE ACQUIRED BOTULISM

- This occurs in older children and adults who acquire botulism through poorly cooked contaminated food with the toxin or through a wound contaminated with soil containing *C. botulinum*.
- Recovery may take months.

3. Peripheral Nerve Disorders

ACUTE INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY

- 🕮 AIDP, also known as GBS, is an acute, primarily demyelinating neuropathy with autoimmune causation.
- It often presents with a prodromal respiratory or GI infection occurring within 1 month of onset. This often occurs with mycoplasma, cytomegalovirus, EBV, *Campylobacter jejuni*, and various vaccinations.
- Weakness usually begins distally in the LEs with progressive ascending paralysis to involve the UEs.
- The most common cranial nerve abnormality is ipsilateral or bilateral LMN facial paralysis.
- Weakness generally peaks within 2 weeks of onset, and time to maximum recovery is approximately 7 ± 5 months. Complete recovery occurs in most children.
- Treatment typically includes plasma exchange and IV immune globulin. Recovery is often good in children without treatment.

CHRONIC INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) has a chronic relapsing course. Electrodiagnostic studies show focal conduction block, temporal dispersion of CMAPs, prolongation of distal motor latencies, markedly slow conduction velocities, and absent or prolonged H-wave and F-wave latencies.

HEREDITARY MOTOR SENSORY NEUROPATHY

HMSN is a group of inherited diseases of peripheral nerves affecting both motor and sensory nerves with progressive neuromuscular impairment. The prevalence is 1 per 2,500, with onset usually in the first or second decade of life. It is the most frequently inherited polyneuropathy:

- **Type I HMSN** (CMT Type I) occurs in 70% to 80% of cases. It presents with a hypertrophic demyelinating neuropathy (onion bulbs) on nerve biopsy. The majority of CMT Type 1 patients demonstrate gene locus of chromosome at 17p11.2–12.
- **Type II HMSN** (CMT Type II) is an axonal neuropathy with low amplitudes noted on nerve conduction studies. It presents at a later age with less involvement of small muscles of the hands. Wasting of the calf and anterior compartment of the leg gives rise to an "inverted champagne bottle" or "stork leg" appearance.
- **Type III HMSN** (Dejerine–Sottas disease) is a severe hypertrophic demyelinating polyneuropathy with onset in infancy or early childhood.
- **Type IV HMSN** (Refsum disease) is an autosomal recessive disorder. Clinical features include distal muscle weakness, impaired sensation, and absent or diminished DTRs. Weakness usually

is present initially in the distal LEs and subsequently in the distal UEs. Progressive weakness may slowly occur more proximally in the knees, elbows, and pelvic and shoulder girdles over decades.

TOXIC NEUROPATHIES

These are rare in North America but more common in other regions of the world.

- *Arsenic*—causes an axonal or demyelinating neuropathy. Diagnosis is from levels in the blood, urine, hair, and nails.
- *Lead*—most commonly occurs secondary to ingestion of lead-based paint. Clinical findings include anorexia, nausea and vomiting, GI disturbance, clumsiness, seizures, mental status changes, and papilledema. Weakness predominantly occurs in the LEs.
- *Mercury*—typically causes a distal motor axonal neuropathy. Occurs from ingestion of mercuric salts, exposure to mercury vapor and/or use of topical ammonia, and mercury ointments. Symptoms may include generalized encephalopathy, fatigue, decreased or absent DTRs, ataxic gait, and often distal sensory paresthesias.
- *Organophosphate poisoning*—this occurs secondary to exposure to insecticides or lubricants from the plastic industry. Symptoms may include confusion and coma (encephalopathy), sweating, abdominal cramps, diarrhea, and constricted pupils, with motor polyneuropathy as a late effect.
- *N-hexane (glue-sniffing) neuropathy*—this occurs in teenaged recreational glue sniffers. It is predominantly a distal motor and sensory demyelinating polyneuropathy.
- *Chemotherapeutic agents, especially vincristine*—this often produces pure motor axonal polyneuropathy. Severity is dose-related. Symptoms include distal weakness, absent DTRs, and often foot drop.

METABOLIC NEUROPATHIES

Most often occur with:

1. End-stage renal disease—predominantly presents as a distal motor and sensory polyneuropathy in a stocking-glove distribution with loss of sensation, loss of vibratory sense, and distal weakness, especially peroneal innervated musculature.
2. Diabetic neuropathy—distal motor and sensory polyneuropathy with both axonal and demyelinating features in stocking-glove distribution. It is less common in children than adults and may be related to degree of glucose control.

4. Motor Neuron Disorders

SPINAL MUSCULAR ATROPHY (TABLE 10–32)

SMA involves a group of inherited disorders characterized by generalized weakness, muscle wasting, and both upper and lower motor neuron signs/symptoms. This is caused by degeneration of both anterior horn cells of the spinal cord (LMN) and brainstem motor nuclei without pyramidal tract involvement (bulbar symptoms). SMA types I to IV are linked to autosomal recessive SMN1 (survival of motor neuron 1) gene mutations on chromosome 5 and are characterized by the time of disease presentation and number of SMN2 (survival of motor neuron 2) copies:

TABLE 10–32 Childhood-Onset Spinal Muscular Atrophy: Clinical Classification

TYPE	ONSET	COURSE	AGE AT DEATH
1 (Severe)	Birth to 6 months	Never sit	Usually <2 years old
2 (Intermediate)	≤18 months	Never stand	>2 years old
3 (Mild)	≥18 months	Stand alone	Adult

Source: From Munsat TL, Davies KE. International SMA Consortium Meeting (26–28 June 1992, Bonn, Germany). *Neuromuscul Disord*. 1992;2(5–6):423–428. doi:10.1016/S0960-8966(06)80015-5, with permission.

  - SMA Type I: At birth to 6 months
  - SMA Type II: 7 to 18 months
  - SMA Type III: After 18 months
  - SMA Type IV: Adulthood
  - The first three types of autosomal recessive SMA are described in more detail in the following section, all of which are linked to chromosome 5q13.
  - X-linked SMA is similar to Type I SMA in its time of onset (early) and disease severity.

With newborn screening and earlier identification of SMA, disease-modifying therapies such as gene therapy onasemnogene abeparvovec-xioi, as well as risdiplam and nusinersen, which both increase SMN protein production, are now treatment options for children with SMA (Day, et al., 2022).

SPINAL MUSCULAR ATROPHY TYPE I: WERDNIG–HOFFMAN DISEASE (SEVERE SPINAL MUSCULAR ATROPHY)

Autosomal recessive disorder of early infancy with severe axial and limb weakness due to degeneration of the anterior horn cell of the spinal cord and brainstem:

- The majority present within the first 2 months of life with generalized hypotonia and symmetrical weakness.
- Symptoms include a weak suck, dysphagia, labored breathing during feeding, frequent aspiration, and a weak cry.
- Examination reveals generalized hypotonia and symmetric weakness of the LEs more than UEs. Proximal muscles are more affected than distal muscles. "Frog leg" position occurs when supine with LEs abducted and externally rotated.
- Diaphragmatic breathing occurs secondary to intercostal and abdominal muscle weakness and relatively preserved diaphragmatic function. Abdominal protrusion, paradoxical thoracic depression, and intercostal retraction are seen. Facial weakness occurs in 50% of cases with tongue fasciculation in 56% to 61%. Preservation of DTRs does not exclude the diagnosis of SMA. Extraocular muscles and myocardium are spared.
- *Age of onset*:
  - In utero or within the first few months of life
- *Presenting symptoms*:
  - Hypotonia and weakness
  - Sucking and swallowing difficulty
  - Respiratory problems
- *Cardinal clinical signs*:
  - Severe limb and axial weakness, frog posture
  - Marked hypotonia
  - Poor head control
  - Diaphragmatic breathing, costal recession
  - Bell-shaped chest
  - Internal rotation of arms, jug-handle posture
  - Legs in frog position
  - Normal facial movements
  - Absent tendon reflexes
  - Weak cry
- *Course and prognosis*:
  - Prognosis poor; majority die of pneumonia in the first year, most within 3 years
  - Prone to respiratory infections
  - Despite severity, weakness usually nonprogressive
- *Investigations*:
  - CK: Normal
  - Ultrasonography: Normal or increased echo plus atrophy of muscle
  - EMG: Motor nerve conduction velocity normal or reduced; poor motor action potential amplitude. Features of denervation on needle EMG.

- *Muscle biopsy*:
  - Large group atrophy plus isolated clusters of large fibers (uniformly Type I)
  - Early cases may show minimal changes (prepathologic).
- *Genetics*:
  - Autosomal recessive, gene 5q11–q13
- *Management*:
  - Pharyngeal suction if bulbar weakness present
  - Spinal brace in less severe cases to maintain sitting posture
  - Supportive treatment of pneumonia (Dubowitz, 1978)

SPINAL MUSCULAR ATROPHY TYPE II: INTERMEDIATE SPINAL MUSCULAR ATROPHY

Autosomal recessive disorder characterized by weakness, predominantly of the legs, with ability to sit unsupported but not to stand due to degeneration of the anterior horn cells of the spinal cord and brainstem.

Progressive kyphoscoliosis and restrictive lung disease are seen in the late first decade of life:

- The disease is slowly progressive with decline of less than one-half of the manual muscle testing unit per decade.
- *Age of onset*:
  - Usually between 6 and 12 months
- *Presenting symptoms*:
  - Weakness of legs
  - Inability to stand or walk
- *Cardinal clinical signs*:
  - Symmetrical weakness of legs, predominantly proximal
  - Able to sit unsupported but unable to stand or take full weight on legs
  - Fasciculation of tongue (about 70%)
  - Tremor of hands
  - Tendon jerks absent or diminished
  - Facial muscles spared
- *Associated features*:
  - Scoliosis
  - Normal or advanced intellect
  - Variable intercostal weakness and respiratory problems
  - Hypotonia and excessive joint laxity, especially hands and feet
- *Course and prognosis*:
  - Muscle weakness usually static and nonprogressive; may show functional improvement—some may have increasing weakness or disability over a long period or during growth spurt or if putting on weight.
  - Long-term prognosis is dependent on respiratory function.
- *Investigations*:
  - CK: Normal or moderately elevated
  - Ultrasonography: Characteristic picture of increased echo in muscle atrophy and increased subcutaneous space
  - EKG: Normal complexes—characteristic baseline tremor, especially in limb leads
  - EMG: Evidence of denervation and reinnervation
- *Muscle biopsy*:
  - Characteristic pattern of large group atrophy plus variable clusters of enlarged fibers, uniformly or predominantly Type I
- *Genetics*:
  - Autosomal recessive, gene 5q11–q13. Alleles of one gene to account for varying severities of SMA, or dual genes or separate genes.
  - 95% of individuals with SMA have monogenic deletions of SMN1 gene with resultant deficiency of SMN protein causing degeneration of motor neurons in the anterior horn of spinal cord.

- *Management*:
  - Prevention of scoliosis by early bracing
  - Treatment of scoliosis by spinal braces or surgery
  - Early achievement of standing posture in standing frame or calipers
- Promotion of ambulation by appropriate orthoses
- *New therapeutic options*:
  - Three FDA-approved disease-modifying agents, all work by restoring SMN protein expression in spinal cord motor neurons:
    - Gene replacement therapy: Onasemnogene Abeparvovec
    - Splicing modifications: Nusinersen, Risdiplam

SPINAL MUSCULAR ATROPHY TYPE III: KUGELBERG–WELANDER SYNDROME (MILD SPINAL MUSCULAR ATROPHY)

Autosomal recessive disorder characterized by proximal weakness, predominantly of the legs, due to degeneration of the anterior horn cells of the spinal cord and brainstem:

- Weakness usually occurs between 18 months and the late teenage years:
  - Proximal weakness occurs with pelvic girdle more affected than shoulder. Hip extensor weakness occurs with increased lumbar lordosis and anterior pelvic tilt.
  - A waddling gait with pelvic drop and lateral trunk lean over stance phase side secondary to hip abductor weakness occurs.
- Fasciculations in the limb muscles and thoracic wall muscles are common.
- Scoliosis is frequent.
- Ventilatory failure secondary to restrictive lung disease is rare.
- *Age of onset*:
  - From the second year of life through childhood and adolescence into adulthood
- *Presenting symptoms*:
  - Difficulty with activities such as running, climbing steps, or jumping
  - Limitation in walking ability—quality or quantity
- *Cardinal clinical signs*:
  - Abnormal gait; waddling, flat-footed, wide base
  - Difficulty rising from floor (+ Gower's sign)
  - Proximal weakness; legs > arms
  - Hand tremor (variable)
  - Tongue fasciculation (variable)
- *Associated features*:
  - Hypermobility of joints, especially hands and feet
- *Course and prognosis*:
  - Weakness usually relatively static; in some may be progressive
  - 🕮 Good long-term survival, depending on respiratory function
- *Investigations*:
  - CK: Normal or moderately elevated
  - Ultrasonography: Characteristic picture of increased muscle echo plus loss of muscle bulk
  - EMG: Motor nerve conduction velocity normal/mildly slowed. Decreased CMAP amplitudes. Evidence of denervation and reinnervation on needle EMG.
- *Muscle biopsy*:
  - Characteristic pattern of large group atrophy plus variable groups of normal or enlarged fibers, often uniformly Type I; or retention of normal bundle architecture with fiber type grouping and focal small group atrophy
- *Genetics*:
  - Autosomal recessive, gene 5q11–q13
  - Less common dominant and X-linked forms
- *Management*:
  - Encourage activity and ambulation

- Rehabilitation and bracing if ambulation lost
- Vigorous treatment of respiratory infections (Dubowitz, 1978)

Spinocerebellar Degeneration Disease

FRIEDREICH'S ATAXIA

- A type of spinal cerebellar degeneration syndrome with an onset before age 20 years
- It is autosomal recessive and linked to chromosome 9q21.
- Symptoms typically start with gait ataxia, which then progresses in an ascending pattern into the rest of the body.
- Other symptoms include weakness/muscle atrophy, dysarthria, gradual loss of proprioception or vibratory sense, absent DTRs, fatigue, nystagmus, and scoliosis.
- The protein abnormality in Friedreich's ataxia is in a mitochondrial protein called frataxin.
- Prevalence of scoliosis approaches 100% with onset in <10 years. Patients usually present with more progressive and severe scoliosis.

EXERCISE IN NEUROMUSCULAR DISEASE

- Eccentric or lengthening contractions produce more mechanical stress on muscle fibers than concentric or shortening contractions.
- No systemic studies using DMD populations have shown any deleterious effect from resistance exercise. Small studies and case series have supported a submaximal strengthening program to increase strength while avoiding muscle damage, exhaustion, and soreness.
- Children with DMD demonstrate lower cardiovascular capacity and peripheral O_2 utilization with a higher resting heart rate compared with controls. Accordingly, aerobic training (i.e., walking, stationary bike, swimming, upper body ergometer) may increase cardiac output and oxygen use by muscles, resulting in decreased fatigue and increased endurance.
- Wheelchair reliance is imminent when knee extension strength becomes less than antigravity (<3/5 strength), and time to ambulate 30 feet is >12 seconds.
- Duration of ambulation in DMD has been successfully prolonged by 2 to 5 years by prompt surgery and bracing following loss of independent ambulation.
- Little evidence supports the efficacy of early prophylactic LE surgery in DMD for independently producing prolonged ambulation.

MANAGEMENT OF SCOLIOSIS IN NEUROMUSCULAR DISEASE (TABLE 10–9)

- Surgical intervention is recommended prior to a curvature of 35 degrees and prior to patient's vital capacity falling to below 35%.
- An FVC of <30% to 40% of predicted normal may contraindicate scoliosis surgical correction secondary to increased perioperative morbidity.
- The management of spinal deformity with spinal orthotics is ineffective in DMD and does not change the natural history of the curve.
- With neuromuscular disease beginning in the first decade of life, spinal bracing is generally used to improve sitting balance in patients unable to walk. Spinal arthrodesis is the most effective treatment of progressive scoliosis.

PULMONARY ISSUES IN NEUROMUSCULAR DISEASE

- 🕮 Pulmonary complications are the number one cause of mortality in childhood neuromuscular diseases.
- Respiratory insufficiency occurs from respiratory muscle weakness and fatigue, alteration of respiratory system mechanics, and impairment of the central control of respiration.

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PAIN MEDICINE

11

Jing Liang, MD • Joseph Lee, MD • Sagar Parikh, MD • Laurent Delavaux, MD • Alexander Shustorovich, DO • Michael Chung, MD

INTRODUCTION

TAXONOMY

- **Acute pain**: Pain lasting for a period of less than 3 months and elicited by injury to tissues for a limited time and stops once the underlying pathology resolves
- **Chronic pain**: Pain lasting for a period of more than 3 months originating from underlying pathology, but symptoms remain past the presumed normal tissue healing process
- **Complex regional pain syndrome (CRPS)**: A chronic pain condition in a limb involving trophic changes, edema, vasomotor changes, and allodynia, typically after an injury or trauma. Symptoms can vary in severity and duration.
  - **CRPS Type I** follows a nonneurologic traumatic injury; formerly known as reflex sympathetic dystrophy (RSD) or Sudeck's atrophy.
  - **CRPS Type II** follows a known traumatic nerve injury; formerly known as **causalgia.**
- **Central pain**: Pain initiating from the central nervous system (CNS)
- **Radiculopathy**: A pathologic process affecting the spinal nerve roots that causes sensory and/or motor dysfunction and/or pain in the innervation of the spinal nerve root(s)
- **Hyperesthesia**: An increased sensitivity to a stimulus
- **Hypoesthesia (or hypesthesia)**: A decreased sensitivity to a stimulus
- **Hyperalgesia**: Exaggerated pain response from a stimulus that normally elicits pain
- **Hypoalgesia**: Diminished sensitivity to a normally painful stimulus
- **Hyperpathia**: An abnormally painful reaction to a stimulus, often seen with a repetitive stimulus or increased threshold
- **Dysesthesia**: An unpleasant sensation that is either evoked or spontaneous in nature
- **Allodynia**: Pain elicited from a nonpainful stimulus
- **Neuralgia**: Pain in a distribution of a nerve or nerves
- **Neuritis**: Inflammation of a nerve or nerves
- **Neurogenic pain**: Pain initiated or caused by a primary lesion or dysfunction in the CNS or peripheral nervous system (PNS)
- **Neuropathic pain**: Chronic pain arising from damage, dysfunction, or injury to a nerve(s)
- **Nociceptive pain**: Pain arising from soft tissue damage (i.e., nonneural injury) or potentially tissue-damaging stimuli that conveys painful information via afferent nociceptive fibers to the CNS
  - **Somatic pain**: A type of nociceptive pain that is carried along a sensory fiber from skin and skeletal musculature
  - **Visceral pain**: A type of nociceptive pain that is carried along a sympathetic fiber from internal organs

- **Nociplastic pain**: Newer term suggested by the international community of pain researchers to describe symptoms with augmented CNS pain and sensory processing and altered pain modulation (nociception)
  - It is a distinct entity separate from neuropathic and nociceptive pain. Peripheral treatments will be less effective (e.g., anti-inflammatory drugs, opioids, surgery, and injections).
  - Mechanisms are incompletely understood, but symptoms are often more widespread, and/or more intense, than would be expected for the level of tissue or nerve damage.
  - It is often mistaken for central sensitization.
  - Often accompanied by fatigue, sleep, memory, and mood problems
- **Paresthesia**: An abnormal sensation whether spontaneous or evoked and includes symptoms such as numbness, tingling and burning
- **Cancer pain**: Pain associated with cancer disease or cancer treatments that may be damaging to the nervous system

ANATOMY, PHYSIOLOGY, AND PHARMACOLOGY OF PAIN TRANSMISSION AND MODULATION

Pain-Signaling Mechanisms

- **Transduction**: Conversion of stimuli to nociceptive electrical impulses. Afferent sensory neurons known as nociceptors transmit pain signals to the CNS. Nociceptor types include mechanical, thermal (heat/cold), and chemical.
- **Transmission**: Electrical activity conducted through the nervous system
  - Peripheral sensory cells → dorsal root ganglion (DRG) → spinal neurons → decussation in spinal cord → brainstem, thalamus, and diencephalic structures
- **Modulation**: Alteration of neural activity (either excitatory or inhibitory) that occurs at the level of the DRG, spinal cord, and supraspinal structures
- **Perception**: Subjective sensation of pain from interplay of neural transmission and modulation, which occurs in somatosensory cortices
- **Nerve Fibers** (see **Figure 11–1A, B**)
  - Majority of pain nerve fibers terminate at Rexed laminae I and-II. The Rexed laminae are divisions within the spinal cord gray matter, named after the neuroanatomist who described them, and separated them based on cellular architecture. Axons from Lissauer's tract enter the dorsal horn gray matter and then branch to connect to neurons within the Rexed laminae (Purves et al., 2001). Laminae I to IV are the main divisions that process pain and make up the dorsal horn of the spinal cord gray matter (Khalid & Tubbs, 2017).
  - 🕮 A-β fibers (mainly tactile, nonpainful fibers) terminate at the spinal cord dorsal horn Rexed laminae III to V.
  - A-δ fibers (fast pain fibers) mainly terminate at Rexed laminae I, III to V.
  - C fibers (slow pain fibers) terminate at Rexed laminae I and II.
- Second-order neurons in the spinal cord are wide dynamic range (WDR) neurons and nociceptive-specific (NS) neurons.
  - 🕮 WDR: Mostly in laminae III to V and receive input from low threshold A-β, nociceptive A-δ (fast pain fibers), and C fibers (slow pain fibers; **Table 11–1**). Responses from WDR are graded, reflecting the type of stimulus fiber.
  - NS: I and II only respond to noxious stimuli.
- 🕮 **Gate Control Theory** (Melzack & Wall, 1965; see also Chapter 8, Physical Modalities, Therapeutic Exercise, Extended Bedrest, and Aging Effects): Input along low-threshold A-β fibers inhibits responses of nociceptive input to WDR cells.
- The anterolateral system and dorsal column-medial lemniscal system carry signals to supraspinal structures.
- Supraspinal modulation occurs in periaqueductal gray (PAG) and rostral ventromedial medulla (RVM) pathway; can be inhibitory or excitatory.
- Cortical structures: Primary and secondary- cortices, insula, anterior cingular cortex, prefrontal cortex, amygdala, and thalamic nuclei

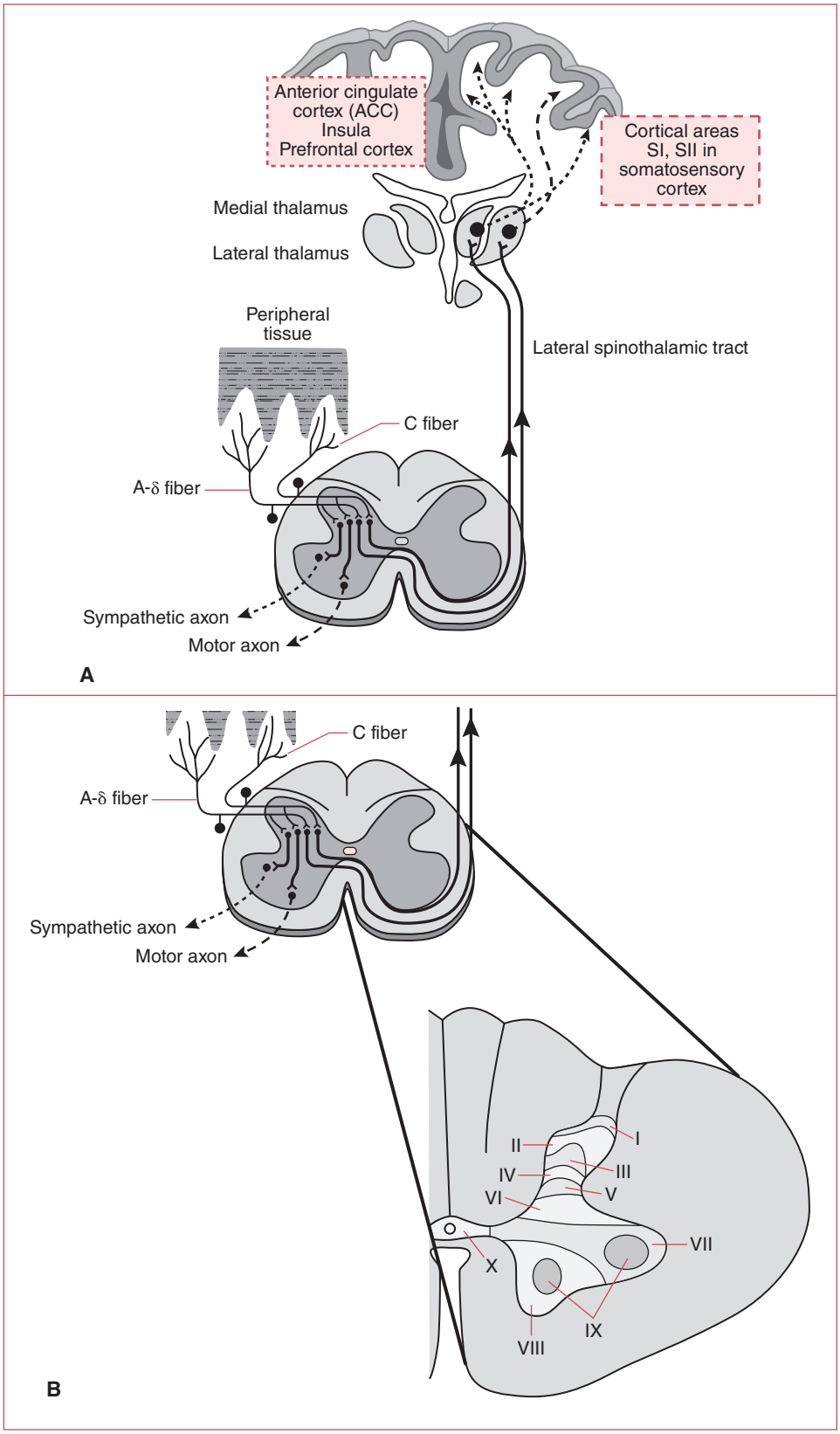


FIGURE 11–1 (A) Pain signaling mechanism. (B) Zoomed-in view of Rexed laminae divisions within the spinal cord gray matter.

TABLE 11–1 Peripheral Sensory Fiber Types

SENSORY FIBER	SIZE (MICRONS)	VELOCITY (M/S)	FUNCTION
A-β	12–14 (myelinated)	30–60	Touch, pressure, vibration, proprioception
A-δ	6–8 (myelinated)	10–15	Sharp pain, light touch, temp
C	<1 (unmyelinated)	<1.5	Dull/achy/burning pain, temp

PHARMACOLOGY

OPIOID PHARMACOLOGY (TABLE 11–2)

TABLE 11–2 Types of Opioid Analgesics

NATURAL	SEMISYNTHETIC	FULLY SYNTHETIC
Morphine, codeine	Hydromorphone, hydrocodone, oxycodone, oxymorphone, diacetylmorphine (heroin), buprenorphine	Fentanyl, methadone, tramadol, dextropropoxyphene

Mechanisms of Action

- Opioids bind to μ (mu), κ (kappa), and δ (delta) in the CNS and peripheral tissues. They presynaptically lower the influx of calcium to reduce neurotransmitter release in sensory C fibers and postsynaptically increase the transport of potassium in the cell to facilitate hyperpolarization in second-order neurons.
- Opioids activate the chemoreceptor trigger zone in the medulla, which leads to gastrointestinal (GI) side effects of nausea and vomiting.
- 🕮 Opioid receptor types (Table 11–3):
  - Mu_1: Analgesia
  - Mu_2: Respiratory depression, sedation, vomiting, euphoria, anorexia, physical dependence, pruritus
  - Delta: Analgesia, spinal analgesia
  - Kappa: Analgesia, sedation, psychotomimetic effects, dyspnea, respiratory depression, euphoria, dysphoria
- 🕮 pKa = pH at which 50% of the drug is ionized. The nonionized form crosses lipid membranes easily. The lower the pKa, the faster the onset.
- 🕮 Octanol/water partition coefficient indicates lipid solubility. Lipid-soluble drugs such as fentanyl exert most of their effect at the localized area where they are administered (less spreading of medication; Figure 11–2; Table 11–4).

🕮 Opioid Metabolites (Figure 11–2)

- Each opioid is metabolized into unique "active" or "inactive" compounds as part of the opioid cascade.
- Any particular compound within the opioid metabolic cascade may be physiologically active, may have different potency than their parent compound, and can often be detected on targeted drug screens.
- **Codeine** and **morphine** are naturally occurring alkaloids from the opium poppy seed, while oxycodone and heroin are semisynthetic compounds.

TABLE 11–3 Opioid Receptor Selectivity

OPIOID	MU RECEPTOR	KAPPA RECEPTOR	DELTA RECEPTOR
ENDOGENOUS OPIOIDS			
Leu—Enkephalin	Low	None	High
Beta—Endorphin	High	High	High
Dynorphin A and B	Moderate	High	Low
OPIOID AGONISTS			
Morphine	High	Low	Low
Diamorphine (heroin)	High	Low	Low
Fentanyl	High	Low	None
Meperidine	High	Low	Low
Methadone	High	Low	High
OPIOID ANTAGONISTS			
Naloxone	High	Moderate	Moderate
Naltrexone	High	Moderate	Moderate
PARTIAL OPIOID AGONISTS			
Buprenorphine	Moderate	Low	None
Pentazocine	None	Moderate	None

- **Morphine** is predominantly metabolized into morphine 3-glucuronide (M3G). A smaller proportion of morphine is metabolized into morphine 6-glucuronide (M6G), a potent opioid agonist, and an even smaller portion is metabolized into hydromorphone (Dilaudid).
- **Codeine** is metabolized into morphine as well as hydrocodone, both of which metabolize into hydromorphone. Hydrocodone also metabolizes into dihydrocodeine.
- **Oxycodone** is metabolized into oxymorphone and hydrocodone.
- Heroin is completely metabolized into 6-monoacetylmorphine (6-MAM), which is then entirely metabolized into morphine.

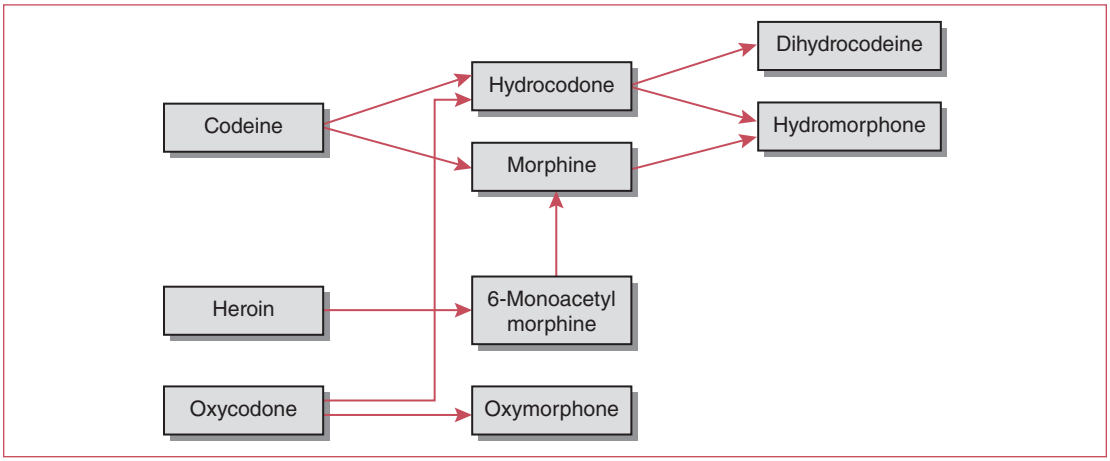


FIGURE 11–2 Opioid metabolites.

TABLE 11–4 Opioid Agonists

OPIOID	pKa	[a]OCTANOL/H_2O PARTITION COEFFICIENT	MECHANISM	METABOLITES	NOTES
Oxycodone	8.5	0.7	Mu agonist	Oxymorphone Noroxycodone	
Oxymorphone	9.3	0.98	Opioid agonist	6-Hydroxy-oxymorphone	
Hydromorphone	8.1	1.28	Mu agonist	Hydromorphone-3-glucuronide	
Tramadol	9.4	1.35	Weak mu agonist, weak NE/5HT reuptake inhibitor	*O*-desmethyltramadol	Contraindicated with MAOI or SSRI (serotonin syndrome)
Morphine	8.0	1.4	Mu agonist	Morphine-3-glucuronide Morphine-6-glucuronide Hydromorphone	Do not use in renal failure
Tapentadol	9.34–10.45	2.87	Mu agonist and NE reuptake inhibitor	Tapentadol-*O*-glucuronide	Do not use in severe hepatic dysfunction
Meperidine	8.5	39	Weak mu agonist	Normeperidine	Do not administer with MAOI
Codeine	8.2	—	Weak mu and delta agonist	Morphine Hydrocodone Codeine-6-glucuronide	Antitussive Antidiarrhea Metabolized to morphine for analgesia
Hydrocodone	8.9	—	Mu and kappa agonist	Dihydrocodeine Hydromorphone	
Methadone	9.3	116	Mu and delta agonist NE and 5HT reuptake inhibitor NMDA antagonist	EDDP (inactive)	May cause QT prolongation
Fentanyl	8.4	860	Mu agonist	Norfentanyl	

[a]Octanol/H_2O partition coefficient: The higher the number, the more lipophilic (less hydrophilic).
5HT, serotonin; EDDP, 2-ethylidene-1,5-dimethyl-3,3-diphenylpyrrolidine; MAOI, monoamine oxidase inhibitor; NE, norepinephrine; NMDA, *N*-methyl-D-aspartate; SSRI, selective serotonin reuptake inhibitor.

🕮 Mixed Opioid Agonist-Antagonists

- **Buprenorphine** (Subutex or Suboxone): Partial mu and kappa agonist, delta antagonist. Suboxone contains naloxone (receptor antagonist) as an abuse deterrent.
- **Pentazocine** (Talwin): Kappa agonist, mu antagonist
- **Butorphanol** (Stadol): Kappa agonist, mu antagonist
- **Nalbuphine** (Nubain): Kappa agonist, mu antagonist

Opioid Side Effects

- Opioids act on the brainstem to reduce sensitivity of the respiratory centers to PCO_2, resulting in respiratory sedation that may progress to death.
- Inhibitory effect on the brainstem nuclei in the cough reflex pathway that suppresses coughing (common use for codeine)
- Constipation: Maintained contraction of the smooth muscles in the gut → decreased motility
- 🕮 Sexual dysfunction (erectile dysfunction, decreased libido): Correlated to hypogonadism and associated with lower testosterone levels

NONOPIOID ANALGESICS

Acetaminophen

- Antipyretic and analgesic: It inhibits prostaglandin synthesis within the CNS (crosses blood–brain barrier) but not peripherally.
- No effects on platelet function or gastric mucosa
- Acetaminophen is available in parenteral form. Propacetamol is administered and then hydrolyzed to paracetamol (acetaminophen). Propacetamol is not available in the United States.
- Be aware of hepatotoxicity (<4 g/day max recommended)

Nonsteroidal Anti-Inflammatory Drugs

- Administered orally, intravenously, intramuscularly, rectally, and topically
- 🕮 Nonsteroidal anti-inflammatory drugs (NSAIDs) inhibit COX-1 and COX-2 enzymes, which produce prostaglandins involved in promoting inflammatory responses after tissue injury. Some NSAIDs display more COX-2 selectivity than others, reducing GI side effects (**Figure 11–3**; **Table 11–5**).

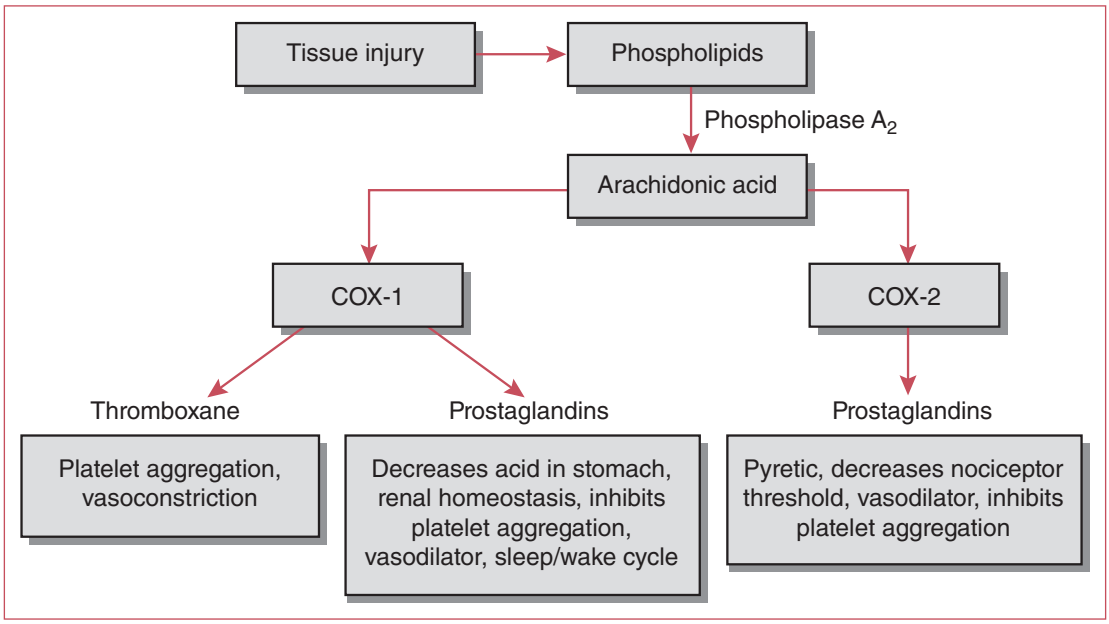


FIGURE 11–3 🕮 Inflammatory pathways.

TABLE 11–5 COX-2 Inhibition Selectivity in NSAIDs

MEDICATION	LESS COX-2 SELECTIVITY	MORE COX-2 SELECTIVITY
Meloxicam		XXX
Celecoxib		XXX
Diclofenac		X
Sulindac (Most common to cause liver failure)		X
Ibuprofen	X	
Naproxen	X	
Salicylate	X	
Indomethacin	XX	
Ketorolac	XXX	

NSAID, nonsteroidal anti-inflammatory drug.
Source: From Warner TD, Giuliano F, Vojnovic I, et al. Nonsteriod drug selectivities for cyclo-oxygenase-1 rather than cyclooxygenase-2 are associated with human gastrointestinal toxicity: a full in vitro analysis. *Proc Natl Acad Sci U S A*. 1999;96(13):7563–7568 with permission.

Antidepressants

- **Tricyclic antidepressants (TCAs):** TCAs have shown analgesic properties, independent from their antidepressant effects. These are mediated by several mechanisms.
  - Inhibit serotonergic and noradrenergic reuptake, acting at the level of the brainstem on the pain-suppressing mechanism of the descending bulbospinal tracts
  - Alter NMDA (*N*-methyl-D-aspartate) receptors and binding characteristics, which may contribute to spinal analgesia and modulation of central sensitization states
  - Modify opioid receptor densities, block sodium channels (especially amitriptyline), and increase densities of L-type calcium channels
  - Undergo hepatic metabolism and clearance through cytochrome P450 enzyme pathway
  - Side effects of arrhythmias with QT_c prolongation may occur. Caution with TCAs given concern for lethal toxicity → the leading cause of overdose death. However, monitoring of TCA serum levels has not been recommended. Do not prescribe in conjunction with selective serotonin reuptake inhibitors, as these compete for hepatic P450 enzyme and can increase TCA plasma levels.
  - Anticholinergic and antihistaminic side effects often dictate patient tolerance.
  - Examples: Amitriptyline, desipramine, nortriptyline, imipramine, doxepin, and clomipramine. Nortriptyline is an active metabolite of amitriptyline with a lower incidence of side effects. Imipramine metabolizes to desipramine.
- **Selective Serotonin Reuptake Inhibitors (SSRIs):** SSRIs inhibit presynaptic serotonin reuptake in the CNS. They have been found to be beneficial for treating depression that may secondarily assist in pain management. Evidence to support analgesia with SSRIs is lacking. These medications are metabolized hepatically.
  - Examples: Citalopram, fluoxetine, paroxetine, and sertraline
- **Serotonin–Norepinephrine Reuptake Inhibitors (SNRIs):** These inhibit serotonin and norepinephrine (NE) reuptake.
  - Examples: Venlafaxine and duloxetine
  - Duloxetine is approved in the United States for diabetic peripheral neuropathic pain, fibromyalgia, generalized anxiety, and major depression. It is the only approved medication for both painful and psychiatric conditions. Venlafaxine can cause hypertension.

  - 🕮 Use with caution: SSRIs or SNRIs given in combination with each other or with TCAs, monoamine oxidase inhibitors (MAOIs), triptans, tramadol, or some antiemetics may result in serotonin syndrome.

Anti-Epileptic Drugs (Membrane-Stabilizing Agents)

- 🕮 **Gabapentin** (Neurontin®): A gamma-aminobutyric acid (GABA) analogue that blocks α2δ subunit of L-type calcium channels and is used to treat neuropathic pain, including from diabetic neuropathy, postherpetic neuralgia, HIV-associated neuropathy, Guillain–Barré syndrome, phantom limb pain, spinal cord injury (SCI) pain, and cancer-associated neuropathic pain.
  - Side effects: Somnolence, dizziness, edema
- **Pregabalin** (Lyrica®): A GABA analogue that blocks α2δ subunit of L-type calcium channels. It requires less frequent dosing than gabapentin (has more linear pharmakinetics than gabapentin). It is used in postherpetic neuralgia, diabetic neuropathy, SCI pain, and fibromyalgia.
  - Side effects: Somnolence, dizziness, dry mouth, edema
- **Valproic acid** (Depakote®) increases GABA effects and may inhibit NMDA/glutamate receptors. It is used for anti-manic, antidepressant, impulsivity, seizures, and migraine prophylaxis.
  - Side effects include thrombocytopenia, neutropenia, anemia, dizziness, sedation, hepatitis, and pancreatitis. Complete blood count (CBC) and liver function tests should be obtained prior to initiation.
- **Lamotrigine** (Lamictal®) inhibits voltage-dependent sodium channels, although the exact mechanism is unknown. It is used in seizure, mood stabilization, and migraine prophylaxis.
  - Side effects: Rash, toxic epidermal necrolysis, dizziness, malaise, headaches (HAs), and ataxia
- **Carbamazepine** (Tegretol®) inhibits voltage-gated sodium channels. Need to monitor level closely in the beginning due to autoinduction. Used in seizures, as a mood stabilizer, and in trigeminal neuralgia.
  - Side effects: Rash, aplastic anemia, and agranulocytosis. CBC should be done every 2 to 4 months.
- **Oxcarbazepine** (Trileptal®) inhibits voltage-gated sodium channels. Use in seizures and trigeminal neuralgia. Has a more favorable side effect profile to carbamazepine.
  - Side effects: Sedation, hyponatremia, dizziness, somnolence, nausea, and vomiting

Skeletal Muscle Relaxants

- This class of medications is typically used in the acute phase for muscle pain and spasm. They were only shown to be effective in certain populations of patients for muscle pain in the first 4 days. No functional improvement has been noted after 1 week, and chronic use is not recommended. These all have additive effects with alcohol, opioids, and other CNS depressants. All medications in this class are metabolized in the liver.
- **Cyclobenzaprine:** While the exact mechanism is unknown, it likely inhibits NE reuptake in locus coeruleus, causing descending inhibition of alpha motor neuron activity and reduction of spinal reflexes. It is used for cervical and lumbar pain and muscle spasms.
  - Side effects: Dry mouth, drowsiness, HA, nausea, diarrhea, dizziness, confusion
- **Carisoprodol:** A central acting muscle relaxant that blocks interneuronal activity in the descending reticular formation and dorsal horn of the spinal cord to depress polysynaptic reflexes. It is used for acute musculoskeletal (MSK) pain.
  - Side effects: Drowsiness, dizziness, HA, hepatotoxicity, ataxia, nausea/vomiting
  - Use with caution: Abuse potential due to meprobamate, its primary metabolite, which is a controlled substance
- **Tizanidine:** Centrally acting, binding to α2-adrenergic receptors, reducing the release of presynaptic neurotransmitters. It is used for spasticity, a paravertebral muscle spasm.
  - Side effects: Dry mouth, drowsiness, HA, dizziness, hypotension, confusion

- **Baclofen:** Activates GABA-B receptors in the brain, reducing excitatory neurotransmitter release in both the brain and spinal cord. There is strong evidence for its use in spasticity.
  - Side effects: Nausea, weakness, drowsiness, confusion, HA, hypotension, weight gain

PAIN SYNDROMES

TABLE 11–6 📖 Types of Pain

	NOCICEPTIVE/SOMATIC PAIN	VISCERAL PAIN	NEUROPATHIC/CENTRAL PAIN
Description	• Deep somatic pain: Dull/aching • Superficial somatic pain: Sharp, pricking • Burning, localized, reproducible	• Crampy and dull • Vague in location	• Burning, tingling, shooting, stabbing, electric-like • May be associated with numbness, tingling
Causes	• Noxious perception from tissue damage can originate from the skin, muscle, bone, or fascia • Mediated by somatic nervous system	• Internal structures of solid or hollow organs/autonomic nervous system; gastrointestinal • Mediated by ANS	• Primary lesion or dysfunction of the pain-sensing nervous system (CNS or PNS)

ANS, autonomic nervous system; CNS, central nervous system; PNS, peripheral nervous system.

MYOFASCIAL PAIN

- Localized and referred soft tissue pain, arising from myofascial trigger points, that can exhibit sensory, motor, and autonomic symptoms. Myofascial pain syndrome can be due to a combination of biomechanical, neurologic, psychological, nutritional, and hormonal components and can lead to a degree of central sensitization with chronic pain.
- Diagnosis is based on a detailed history (including functional status) and physical examination with specific clinical characteristics.
  - Most often caused or worsened by poor biomechanics and/or posture, leading to repetitive microtrauma and decreased range of motion (ROM) of the muscle tissue. There will often be a distinct distribution of regional pain and stiffness, worsening with muscle overload or repetitive activities. In some cases, there is a more central pain generator as well.
  - Presence of trigger point(s) with a consistent reproducible referred pain pattern. A myofascial trigger point is a hyperirritable taut band of skeletal muscle or fascia that is sensitive and painful to digital compression. Palpation or needling of a trigger point may elicit an involuntary focal muscle contraction called the twitch response.
- 📖 Treatments:
  - Physical therapeutics, modalities (heat/ice, ultrasound, transcutaneous electrical nerve stimulation [TENS]), cooling analgesic spray, stretch, massage, postural alignment, acupressure (ischemic compression therapy)
  - Trigger point injection with or without injectants (dry needling). Injectants can include local anesthetics as well as sterile water/saline. Injections with corticosteroids have not been shown to be effective in treating myofascial pain. Botulinum toxin has been used in significant, refractory cases. Postprocedural stretching is important.

HEADACHES (TABLE 11–7)

📖 Migraine Headaches

- Presentation:
  - Symptoms: Unilateral > bilateral, pulsing/throbbing, stabbing, frontal, or temporal pain. It can occur with or without aura.

TABLE 11–7 Categories of Headaches

	TYPES OF HA	ETIOLOGY
Primary HAs	Migraine HA Tension HA Cluster HA Hemicrania continua	No apparent underlying organic disease etiology
Secondary HAs	HAs are considered a symptom of an underlying disease	Caused by an underlying organic process

HA, headache.

  - Can be associated with nausea, vomiting, photophobia, phonophobia, vertigo, diarrhea, or diaphoresis
  - Typically last for 30 minutes to 1 day but can also be intractable and last up to 1 week (status migrainosus)
- Two migraine subtypes:
  - Migraine without aura (common)
  - Migraine with aura (classic): Focal neurologic symptoms preceding or accompanying HAs
    - Focal neurologic symptoms: Visual disturbances (bright spots, dark spots, tunnel vision), tremor, pallor, vertigo, unilateral numbness or weakness, transient aphasia, or thick speech that precedes the HA
    - Premonitory and resolution symptoms: Hyperactivity, hypoactivity, depression, craving for particular foods, repetitive yawning, and other less typical symptoms are reported by some patients.
- **Cortical spreading depression (CSD) theory:** A phenomenon defined as a wave of neuronal depolarization in the cortical gray matter followed by neuronal activity suppression, resulting in blood flow changes (hyperemia, then oligemia)
- **Neurovascular theory:**
  - Trigeminovascular system: Plexus of unmyelinated fibers from trigeminal (cranial nerve [CN] V) ganglion innervating cerebral and pial arteries, venous sinuses, and dura mater.
  - Stimulation of trigeminal ganglion by CSD causes antidromic activation of the trigeminovascular system, resulting in release of peptide neurotransmitters: Substance P, neurokinin A, and calcitonin gene-related peptide (CGRP).
  - 🕮 These peptide neurotransmitters vasodilate nearby blood vessels, which causes extravasation of plasma or "sterile neurogenic inflammation." This stimulates the trigeminal nerve endings, causing nociceptive orthodromic activation to the trigeminal ganglion, resulting in pain.
- **Vascular theory:** Intracranial vasoconstriction results in ischemia, which then causes the aura that precedes migraines. Subsequent rebound vasodilation and activation of perivascular nociceptors result in migraine HA.
- **Treatments (Table 11–8):**
  - A stratified care approach uses **Mi**graine **D**isability **As**sessment (MIDAS) scores to choose treatment based on severity and degree of disability.
  - "Stepped care" approach: Simple analgesics first, then migraine-specific medications
  - Alternative methods: Botulinum toxin injections, cognitive behavioral therapy (CBT), biofeedback, and relaxation techniques

Tension-Type Headaches

- 🕮 Most common HA type
- Presentation:
  - Mild-moderate, bilateral "tight band" or pressure pain around the head. Tends to be less intense and less localized than migraine HA.
  - Usually lasts from 30 minutes to several days or weeks
- Treatment: Over-the-counter (OTC) analgesics (aspirin, acetaminophen, or ibuprofen)

TABLE 11–8 Medications for Migraine Headaches

ABORTIVE MEDICATIONS	PREVENTIVE MEDICATIONS
1. Acetaminophen 2. NSAIDs (e.g., aspirin, ibuprofen, naproxen, ketorolac) 3. Opioids (butalbital, meperidine) 4. Adjunctive medications (prochlorperazine and metoclopramide) 5. Ergotamines (FDA pregnancy category X) 6. Triptans (5-HT 1B and 1D agonists)	1. Beta-blockers (e.g., propranolol, timolol) 2. NSAIDs 3. TCAs (e.g., amitriptyline) 4. SSRIs (e.g., fluoxetine) 5. Antiepileptics (valproic acid, carbamazepine, gabapentin) 6. Magnesium 7. Botulinum toxin injections

5-HT, 5-hydroxytryptamine; NSAIDs, nonsteroidal anti-inflammatory drugs; SSRIs, selective serotonin reuptake inhibitors; TCAs, tricyclic antidepressants; U.S. FDA, U.S. Food and Drug Administration.

Cluster Headaches

- Presentation:
  - Episodic, severe, stabbing, unilateral, periorbital/temporal HA pain. Periorbital pain affecting the distribution of the first trigeminal nerve (temple, forehead, cheek).
  - It can be associated with ipsilateral autonomic symptoms—miosis, ptosis, conjunctival injection, lacrimation, and rhinorrhea.
  - Pain lasts 15 to 90 minutes and can occur up to eight times per day (in clusters).
  - Striking feature is the daily circadian rhythm (occurs around the same time every day) and certain seasons as well
  - Usually occur between the ages of 20 and 40 and predominantly affects males
  - Typically considered unresponsive to indomethacin, although there are documented case reports of indomethacin-responsive cluster HAs
- Treatment:
  - Prophylactic: Verapamil, methysergide, steroids, ergotamine, dihydroergotamine (DHE), lithium, and antiepileptics
  - Abortive: 100% oxygen (7–12 L), sumatriptan, DHE, intranasal lidocaine 4% to 6%

HEMICRANIA CONTINUA (Bahra, 2023)

- Continuous unilateral HA with superimposed exacerbations and autonomic symptoms
- Similar symptoms as cluster HAs
- Unlike cluster HAs, hemicrania responds to indomethacin.

PAROXYSMAL HEMICRANIA (Bahra, 2023)

- Similar to cluster HA but occurs more frequently (15–20× daily) with shorter duration (5–45 minutes)
- Unlike cluster HAs, paroxysmal hemicrania also responds to indomethacin.

CANCER PAIN

- Both nociceptive and neuropathic pain can be somatic, visceral, central pain, or a combination of these.
- **Multiple myeloma:** Pain may be a result of a bone fracture or a tumor mass effect on a nerve. Radiation therapy and surgery can control myeloma pain by shrinking tumors that are compressing nerves.
- **Pancreatic cancer:** Severe upper abdominal and back pain is a significant complication in patients who develop pancreatic cancer. For pain unresponsive to other measures, celiac plexus blocks, external beam radiation, and intrathecal pain pumps may help.
- **Ovarian cancer:** In its earliest stages, it can cause mild to no symptoms. Patients may present with abdominal or pelvic pain that may extend into the back and legs. In its advanced stages, pain is more common.
- **Prostate cancer:** Most commonly diagnosed cancer in American men and the second leading cause of cancer death. Many experience pelvic pain. The majority of patients with advanced

disease have sclerotic bone metastasis that can cause mild to very severe pain. Enlargement and inflammation of the primary prostatic tumor may also cause urethral, rectal, suprapubic, and penile pain.

- **Breast cancer**
  - 🕮 Postmastectomy pain syndrome: Intercostobrachial neuralgia (T1–T2). Chronic neuropathic pain that develops shortly after or several months post surgery. Prevalence is higher after lumpectomy than mastectomy and especially when axillary lymph node dissection has been performed. Pain is localized to the axilla, shoulder, arm, and/or chest wall.
  - Lymphedema: Usually develops within 2 years after breast cancer surgery. Risk dependent upon the type of surgery, the time since surgery, and if radiation therapy were used. Causes a heavy sensation or aching discomfort in the arm, swelling of the affected arm/upper chest, and/or numbness/tingling of the arm along with fatigue. Recommended treatment is a lymphedema therapy program combining massage, skin care, exercise, and compression garments.
  - Pain after radiation therapy: During the acute treatment period, radiotherapy can cause painful skin irritation ranging from mild erythema to significant moist desquamation. The skin may peel and weep like a sunburn. There have been reported cases of long-term reactions, such as cervical or brachial radiation-induced plexopathies (RIP).
  - Pain after chemotherapy: Anthracyclines and alkylating agents can cause oral mucositis (painful mouth ulcerations). Acute paclitaxel syndrome consists of arthralgias and myalgias, which range from mild to debilitating. Taxanes can cause painful peripheral neuropathy in up to 60% of patients.
- **Lung cancer:** Three main causes of pain associated with advanced lung cancer are skeletal metastatic disease (34%), Pancoast tumor (31%), and chest wall disease (21%).
  - Chest pain worsens with disease progression; this is prevalent in approximately 20% of patients with lung cancer. Pain is frequently at ipsilateral chest as the tumor site. Often requires radiotherapy to alleviate the pain.
  - Costopleural syndrome: Severe, intractable chest pain that is a common symptom in pleural mesothelioma as it invades the pleural cavity and thoracic wall. Characteristically pleuritic, lateralized, dull, or diffuse; neuropathic components present secondary to entrapment of thoracic, autonomic, or brachial plexus nerves. Can progress relentlessly during the course of the cancer and is hard to control. Percutaneous cervical cordotomy has been shown to relieve pain. The procedure interrupts the spinothalamic tract at C1/C2, causing contralateral loss of pain perception below the level of the lesion.
  - Metastasis: Liver metastases frequently cause right upper quadrant pain. Brain metastases from non-small cell lung cancer occur in approximately 33% of patients and may cause HA.
  - 🕮 Hypertrophic pulmonary osteoarthropathy: Clubbing and periosteal proliferation of the tubular bones, causing a symmetrical painful arthropathy affecting the ankles, knees, wrists, and elbows. Pain improves if the tumor is resected. In advanced lung cancer, NSAIDs/bisphosphonates are treatment mainstays.

World Health Organization Analgesic Ladder for Cancer Pain Relief (See Also Figure 9–11)

- Mild to moderate pain: Nonopioid analgesics ± adjuvant
- Moderate pain: Short-acting opioids ± nonopioid analgesics ± adjuvant
- Moderate to severe pain: Short- and long-acting opioids ± nonopioid analgesics ± adjuvant

Invasive Approaches to Cancer Pain

- **Intraspinal administration of opioid analgesics** through epidural, intrathecal, or intraventricular routes (for both neuropathic and nociceptive pain relief)

- **Neuroablative techniques:**
  - Peripheral neurolytic blockade
  - Ganglionic blockade
  - 🕮 Dorsal root entry zone (DREZ) lesioning: A surgical technique that selectively destroys neurons located in the posterolateral spinal cord. It is used to treat medically refractory chronic pain syndromes associated with neurons that develop paroxysmal hyperactivity following deafferentation injury (e.g., brachial plexus avulsion).
  - 🕮 Cordotomy: A surgical procedure that ablates the spinothalamic tract, providing selective loss of pain and temperature perception several segments below and contralateral to where the lesion is placed. Used for severe pain secondary to cancer (especially for pleural and peritoneal mesothelioma) where treatment to level 3 of the World Health Organization's (WHO) pain ladder is ineffective. Bilateral cordotomies may be required for visceral or bilateral pain. The effect is usually temporary, so it is typically used in patients with a short life expectancy.
  - Punctate midline myelotomy: A neuroablative procedure that interrupts the midline of the dorsal column (which contains a pathway for nociceptive visceral signals) to treat intractable abdominal and pelvic cancer pain.
  - Cingulotomy: Bilateral anterior cingulotomy targets the anterior cingulate cortex (part of the limbic system). It was used to treat psychiatric disorders such as obsessive-compulsive disorder and depression. It was more recently used for acute cancer pain when other methods failed.

🕮 COMPLEX REGIONAL PAIN SYNDROME

- Also see the "Complex Regional Pain Syndrome Type I" section in Chapter 1, Stroke.
- This neuropathic pain disorder affects one or more extremities and usually occurs following a physical injury. CRPS is believed to be caused by dysfunction of the central and peripheral nervous systems, resulting in allodynia, edema, changes in skin blood flow, and/or abnormal sudomotor activity (**Table 11–9**).

TABLE 11–9 Revised CRPS Criteria Adopted by the IASP in 2012

General Features of the Syndrome:
CRPS is a syndrome characterized by a continuing (spontaneous and/or evoked) regional pain that is seemingly disproportionate in time or degree to the usual course of any known trauma or lesion. The pain is regional (not in a specific nerve territory or dermatome) and usually has a distal predominance of abnormal sensory, motor, sudomotor, vasomotor, and/or trophic findings. The syndrome shows variable progression over time.

***Clinical* Diagnostic Criteria for CRPS**

1. Continuing pain, disproportionate to any inciting event
2. Must report at least one symptom in *three of the four* following categories:
   - ***Sensory***: Reports of hyperalgesia and/or allodynia
   - ***Vasomotor***: Reports of temperature asymmetry and/or skin color changes and/or skin color asymmetry
   - ***Sudomotor/Edema***: Reports of edema and/or sweating changes and/or sweating asymmetry
   - ***Motor/Trophic***: Reports of decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair, nail, skin)
3. Must display at least one sign[a] at time of evaluation in *two or more* of the following categories:
   - ***Sensory***: Evidence of hyperalgesia (to pinprick) and/or allodynia (to light touch and/or deep somatic pressure and/or joint movement)
   - ***Vasomotor***: Evidence of temperature asymmetry and/or skin color changes and/or asymmetry
   - ***Sudomotor/Edema***: Evidence of edema and/or sweating changes and/or sweating asymmetry
   - ***Motor/Trophic***: Evidence of decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair, nail, skin)
4. There is no other diagnosis that better explains the signs and symptoms

[a]A sign is counted only if it is observed at time of diagnosis.
CRPS, complex regional pain syndrome; IASP, International Association for the Study of Pain.
Source: From Harden RN, McCabe CS, Goebel A, et al. Complex regional pain syndrome: practical diagnostic and treatment guidelines, 5th edition. *Pain Med*. 2022;23(suppl 1):S1–S53. doi:10.1093/pm/pnac046. PMID: 35687369.

- **Two main types:**
  - **CRPS I** (formerly RSD) is diagnosed if the previous symptoms occur from soft tissue injury without confirmed nerve injury in the affected limb.
  - **CRPS II** is diagnosed if the symptoms occur after direct nerve damage. Also known as causalgia.
- Treatments:
  - Functional restoration in a multidisciplinary setting is a hallmark of successful management.
  - Medications: NSAIDs, antidepressants, anticonvulsants, opioids (limited data), topical dimethyl sulfoxide (DMSO), and oral *N*-acetylcysteine (NAC)
  - Therapeutic sympatheticAnerve blocks may be useful if diagnostic sympathetic blocks are positive. Case reports support use of intrathecal baclofen and spinal cord stimulation (SCS). Intravenous (IV) lidocaine infusion is used in refractory cases.

CHRONIC PELVIC PAIN

- **Common gynecologic etiologies:** Ovarian cysts, primary and secondary dysmenorrhea, pelvic congestion, and endometriosis. Pelvic laparoscopy/laparotomy to remove endometrial implants and lyse adhesions has been shown to improve pelvic pain for at least 1 year in approximately 67% of women.
- 🕮 **Interstitial cystitis (painful bladder syndrome):** Chronic inflammatory condition of the bladder's submucosal and muscular layers. Women > men. Bladder wall becomes irritated and can become scarred or stiff.
  - Glomerulations, petechial hemorrhages, on the bladder wall are seen; research questions whether these are caused by cystoscopic hydrodistention.
  - Hunner's ulcers, patches of broken skin on the bladder wall, are present in 10% of interstitial cystitis patients.
  - Symptoms: Often misdiagnosed as a cystitis or urinary tract infection, as a patient can experience dysuria, urinary urgency and frequency, nocturia, and painful sexual intercourse.
  - Diagnosis: Diagnosis of exclusion, cystoscopy, hydrodistention, potassium sensitivity test, urodynamics evaluation
    - Potassium sensitivity test: K+ and water are placed into the bladder at one time. Test is positive if the patient experiences more pain/urgency with K+ and water than with water alone.
  - 2011 American Urological Association interstitial cystitis treatment guidelines amended in 2022 (Clemens et al., 2022):
    - Treatment includes a multimodal pain management approach, which should be continually reassessed over time, and varies per patient. Treatment options are listed below, but have varying levels of evidence strength, and each may not be appropriate for any individual patient (see article for full details). They include:

    1. Behavioral treatments: Patient education (including discussion of normal and abnormal bladder function, benefits vs. risks of treatment options, knowledge that no one treatment has been found effective for most patients, and multiple treatment options may need to be tried before finding relief); behavior modifications; stress management; manual physical therapy (PT) techniques (however, pelvic floor strengthening exercises should be avoided).
    2. Oral medications:
       - May include urinary analgesics, acetaminophen, NSAIDs, opioid/nonopioid medications
       - Amitriptyline, cimetidine, hydroxyzine, and pentosan polysulfate (potential risk with pentosan polysulfate for macular damage, vision injury) may also be considered.
    3. Intravesical instillations (DMSO, heparin, lidocaine) may be used.
    4. Treatment of Hunner's ulcers: Fulguration (with laser or electrocautery) and/or triamcinolone injection; oral cyclosporine A may be offered only if refractory to fulguration/triamcinolone.

5. Procedures: Cystoscopy with short-duration, low-pressure distillation; intradetrusor onabotulinumtoxin A, neuromodulation
6. Surgical intervention: Substitution cystoplasty, urinary diversion

- 🕮 **Irritable Bowel Syndrome (IBS):**
  - Symptom-based diagnosis of abdominal pain or discomfort for 6 months and at least three times per month for the last 3 months without other diagnostic cause.
  - Women > men. Age <45 years old.
  - Symptoms: Chronic abdominal pain, discomfort, bloating, urgency to move bowels, tenesmus, alteration of bowel habits (diarrhea-predominant [IBS-D], constipation-dominant [IBS-C], or both [IBS-M]), or improvement of pain with bowel movement. Onset likely to occur after an infection (IBS-PI), stressful life event, or maturity onset.
  - Comorbidities: HA, fibromyalgia, depression, or chronic fatigue syndrome
  - Treatment: Diet modification, psychotherapy, probiotics, oral medications (fiber supplements, laxatives), antidiarrheals (loperamide), antispasmodics (hyoscine, cimetropium, pinaverium), antidepressants (TCAs, SSRIs), or lubiprostone (Amitiza)
- **Neuropathic pelvic pain**
  - 🕮 Anatomy: Ilioinguinal nerve (T12–L1), iliohypogastric (T12–L1), genitofemoral (L1–L2), lateral femoral cutaneous (L2–L3), obturator (L2–L4)
  - Interventional procedures: Sacral nerve block, ilioinguinal block, hypogastric block
    - Sacral nerve block: Sacral nerves provide sensory innervation to the anorectal region and motor innervation to the external anal sphincter and levator ani muscles. S1 to S4 provide the majority of visceral innervation to the bladder, urethra, and external genitalia.
    - Laparoscopy, total abdominal hysterectomy (TAH), presacral neurectomy, laparoscopic uterosacral nerve ablation (LUNA) procedure
    - Presacral neurectomy: Aids in midline chronic dysmenorrhea that is unresponsive to medical management. Total transection of the presacral nerves, superior hypogastric plexus, lying within the boundaries of the interiliac triangle; usually results in 90% relief of pain.
    - LUNA procedure: Using laparoscopy, the uterus is anteverted with a uterine manipulator and the uterosacral ligaments, which carry nerve fibers, are identified and transected close to their cervical attachment. Studies show that long-term pain relief is <50%.
- **Piriformis syndrome:** A controversial diagnosis in which irritation of the sciatic nerve is thought to be caused by the compression from the piriformis muscle. It can result in pain in the posterior hip girdle musculature with numbness and tingling that may radiate distally into the leg. The prevalence of piriformis syndrome is estimated to be approximately 5% (Papadopoulos & Khan, 2004).
  - Diagnosis: A thorough history and physical exam should be performed to rule out more common causes that refer pain in that region, notably the spine and hip.
  - Physical exam findings: FADIR (flexion, adduction, internal rotation of the hip) test, Freiberg maneuvers, Pace sign, and pain with direct palpation of the piriformis, but clinical testing has not proven to be definitive.
  - Diagnostic studies such as imaging of the area (MRI, CT) and electromyography (EMG) are typically normal.
  - Treatment: PT, NSAIDs, analgesics, muscle relaxants. If treatment fails, a piriformis trigger point injection may be considered.
- Other etiologies: Prostatitis, ureteral obstruction, prostadynia, sexual/physical abuse, cancer pain, psychiatric disorders, and surgical adhesions

PAIN ASSOCIATED WITH OTHER MEDICAL CONDITIONS

- **HIV**
  - 50% of HIV-related pain can also be due to symptoms from disease-associated infections (i.e., HA with cryptococcal meningitis, abdominal pain with *Mycobacterium avium* complex [MAC] infection). 15% to 30% are due to medication side effects (zidovudine can cause HA). 25% to 40% are unrelated to HIV or its treatment.

  - Peripheral neuropathic pain is clinically present in approximately 30% of HIV/AIDS individuals; didanosine and stavudine can also cause peripheral neuropathy.
- **Multiple sclerosis (MS)**
  - Almost 50% of individuals experience chronic pain. Pain typically exhibits as MSK pain (most often seen in the hips, legs, arms), spasticity, paroxysmal pain (most characteristic is facial pain of trigeminal neuralgia; tonic spasms), and neurogenic pain (dysesthesias; most common pain syndrome in MS; often occurs in legs; Lhermitte's sign).
  - Optic neuritis is a common first MS symptom. Usually resolves in 7 to 10 days.
- **Peripheral vascular disease (PVD)**
  - Approximately 50% of individuals are asymptomatic.
  - Pain can vary from calf pain on exercise (vascular claudication) to pain at rest (critical limb ischemia). Achiness, fatigue, and burning in the feet, calves, or thighs. Pain and cramps may occur at night.
  - Noticeable skin color changes, diminished distal pulses, skin ulceration, and gangrene
- **Sickle cell anemia**
  - Pain can be acute, chronic, or a combination of the two. If acute, pain tends to be sudden, intense, and unpredictable from a vaso-occlusive crisis (sickle cell crisis). Chronic pain can be caused by avascular necrosis (AVN, aka osteonecrosis) of the bone (hips > shoulders > ankles), including the spine ("fish-mouth" appearance on x-ray), resulting from repeated vaso-occlusive crises. Knees are rarely involved. Pain can also come from infections such as osteomyelitis and pulmonary infections due to impaired immune response in sickle cell patients.
- **Osteoarthritis (OA)**
  - A degenerative joint disease that occurs several years or decades. It is most commonly found in the hands (Heberden/Bouchard nodes), knees, hips, and spine. Pain is usually a deep ache in the joints exacerbated with extensive use. Decreased ROM and crepitus are frequently present. Stiffness may develop during rest; morning stiffness usually lasts for <30 minutes.
- **Rheumatoid arthritis (RA)**
  - An immune-mediated systemic inflammatory disease affecting multiple organ systems as well as the large joints in the body. It is characterized by inflammation of the membranes lining the joints and synovium. Pain is a result of synovitis, usage-related joint pain secondary to weakness, or secondary arthritis. Usual drugs for treatment are NSAIDs, disease-modifying antirheumatic drugs (DMARDs), and corticosteroids. Morning stiffness usually lasts for >30 minutes.
- **Ankylosing spondylitis (AS)**
  - Typically, the first symptoms are frequent pain and stiffness in the sacroiliac (SI) area, lower back, and buttock that come on gradually over a few weeks or months. Pain can be unilateral, bilateral, or alternate between sides. It is usually dull and diffuse. It is typically worse in the morning and night but may improve with a warm shower or light exercise. Pain is typically chronic, affects both sides, and radiates up the spine into the neck. Tenderness can also spread into the ribs, shoulder blades, hips, thighs, and heels (tendonitis). Among rheumatologic diseases, it has the longest average diagnosis delay of 5 to 10 years (Fallahi & Jamshidi, 2015).
- **Fibromyalgia**
  - Second most common rheumatologic disorder behind OA
  - 3:1 Female:male ratio, associated with >50 years of age, low educational level, low socioeconomic status, and living in rural areas
  - Mechanism: CNS sensitization leads to "disordered afferent processing" and increased levels of excitatory neurotransmitters (glutamate, substance P) and/or low levels of inhibitory neurotransmitters (5-hydroxytryptamine [HT], NE, GABA).
  - Core symptoms: Multifocal pain, fatigue, insomnia, cognitive or memory problems, and psychological distress

- Often coexists with regional MSK pain syndromes, chronic fatigue syndrome, IBS, irritable bladder syndrome, interstitial cystitis, HA, vulvodynia, and pelvic pain.
- Diagnosis: See recent updated Fibromyalgia criteria outlined in Chapter 3, "Rheumatology."
- Management is key (Table 11–10).

TABLE 11–10 Management of Fibromyalgia

PHARMACOLOGIC TREATMENTS OF FM	NONPHARMACOLOGIC TREATMENTS OF FM
1. TCAs and cyclobenzaprine shown to be efficacious in RCTs 2. Pregabalin (FDA approved in 2007) 3. Duloxetine (FDA approved in 2008) 4. Milnacipran (FDA approved in 2009)	1. Exercise: RCTs show benefit both symptomatically and in functional capacity 2. Behavioral medicine approaches (CBT, relaxation techniques, biofeedback) 3. Educational approaches

CBT, cognitive behavioral therapy; FDA, U.S. Food and Drug Administration; FM, fibromyalgia; RCTs, randomized controlled trials; TCA, tricyclic antidepressants.

PAIN OF SPINAL ORIGIN

- See also the Spine section in Chapter 4, Musculoskeletal Medicine.

Radicular Pain

- Results from spinal nerve root inflammation, compression, or damage.
- Common causes of nerve root irritation:
  - An intervertebral disc herniation (leakage of the nucleus pulposus [NP] beyond the annulus fibrosis [AF]) can lead to inflammation and compression of the spinal nerve root. This can result in leakage of neurotoxic substances, which leads to elevated levels of phospholipase A2, leukotriene B4, and thromboxane B2. These inflammatory mediators play an important role in the immune and inflammatory reaction involved in discogenic and radicular pain syndromes.
  - Spinal stenosis, synovial facet, or ligamentum flaval cysts. Less common etiologies include infection and malignancy.
- Annular tears and disc herniation (leakage of NP) can lead to inflammatory processes and release of phospholipase A2, which can sensitize nerve endings and generate pain.
- Clinical presentation: Burning, sharp, shooting, and electric-like pain originates from the spinal region and radiates along the affected dermatome. Radicular pain from a symptomatic lumbar disc herniation is typically aggravated by bending, sitting, and coughing, sneezing, or Valsalva maneuver. Radicular pain from spinal stenosis is typically aggravated by standing and walking and relieved with bending forward, using a shopping cart ("shopping cart sign"), and sitting.
- Treatment: PT, modalities, anti-inflammatory meds, oral steroids, neuropathic agents, epidural steroid injections (ESIs), surgery

Facet Syndrome

- Facet joints are true synovial joints that allow the spine to flex, extend, and rotate. They are formed by the articulation of the superior articular process (SAP) of the inferior vertebra with the inferior articular process (IAP) from the superior vertebra and are innervated by medial branches arising from spinal nerve dorsal rami.
- The dorsal ramus of the nerve root is divided into the lateral, intermediate, and medial branches:
  - Lateral branch: Innervates the paraspinal muscles, skin, and SI joint complex.
  - Intermediate branch: Innervates the longissimus muscle.
  - 🕮 Medial branch: Innervates the facet joint, multifidus muscle, interspinal ligament and muscle, and periosteum of the neural arch.
- 🕮 In the lumbar spine, each facet is innervated by the medial branch nerves at the same level and the level above (e.g., L4–L5 facet joint is innervated by the L3 and L4 medial branches).

Note that the L5 dorsal ramus does not have a medial branch nerve and instead innervates the facet joint directly.

- In the cervical and thoracic spines, each facet is innervated by the medial branches of the dorsal rami from the corresponding spinal levels (e.g., C5–C6 facet joint is innervated by the C5 and C6 medial branches).
- The innervation for the C2 to C3 facet joint comes from the dorsal ramus of C3. The C3 dorsal ramus divides into two medial branches, the larger of which is known as the third occipital nerve.
- Clinical presentation: Axial, nonradicular spinal pain. Traditionally, patients with facet syndrome were thought to have facet pain aggravated by extension and rotation ("facet loading"). However, the literature has shown that clinical history, examination, and imaging findings are unreliable in accurately diagnosing facet-mediated pain. Double diagnostic medial branch blocks (MBBs) should be performed to properly diagnose a facet syndrome prior to consideration of therapeutic interventional treatments.
- Treatment:
  - Conservative: PT, medications
  - Interventional: Diagnostic MBBs; facet blocks, radiofrequency ablation (RFA)

Discogenic Pain

- Also see "The Intervertebral Disc" section in Chapter 4, Musculoskeletal Medicine.
- An intervertebral disc is composed of an outer AF layer that surrounds the NP.
- The AF is composed of Type I collagen. The NP is composed of water and proteoglycans in a Type II collagen network. During weight bearing, the NP transmits force outward to the AF and endplates.
- 🕮 Innervation: Posteriorly by sinuvertebral nerves, laterally by ventral rami, anterolaterally by gray rami communicantes.

TABLE 11–11 Dallas Discogram Classification Scale of Internal Disc Disruption

DALLAS GRADE	ANNULAR DISRUPTION
0	No annular disruption
1	NP leaking into inner 1/3 annulus
2	NP leaking into outer 1/3 annulus
3	NP leaking beyond outer annulus

NP, nucleus pulposus.

Vertebrogenic (Discogenic) Pain (Conger et al., 2022)

- Refers to axial low back pain that is related to anterior column pain. It differs from discogenic pain in the sense that etiology is secondary to degenerative changes to the vertebral endplates, which are innervated by the unmyelinated basivertebral nerve.
- Pain is worse with prolonged sitting > standing, often described as deep axial low back pain, not typically with radiation but may have a referred pain pattern and associated radiculopathy. This pain pattern is similar to discogenic pain.
- Characterized by Modic changes on MRI. Diagnosis can potentially be considered when MRI demonstrates the presence of Modic Type 1 or 2 changes, with or without endplate defects, *and* other potential etiologies of axial spinal pain are ruled out.
- 🕮 Modic classification of degenerative vertebral endplate changes:
  - Modic changes are structural vertebral endplate changes seen on MRI. Modic changes were first described and separated into three different types of changes by Dr. Michael Modic in 1988.
  - Of note, while some studies show association of low back pain with Modic changes, there is no consensus in the medical literature of whether Modic changes have a causal relationship with chronic low back pain (Viswanathan et al., 2020).

- 🕮 Modic Classification of Degenerative Vertebral Endplate Changes on MRI (Modic et al., 1988):

MODIC TYPE	CHARACTERISTICS	🕮 MRI CHANGES
Modic Type I (Edema)	Associated with fissured endplates, bone marrow edema, inflammation, and hypervascular fibrous tissue	Bright on T2, dark on T1
Modic Type II (Fatty changes)	Fatty bone marrow replacement	Bright on T2 and T1
Modic Type II (Bony sclerosis)	Rare; indicative of bony sclerosis	Dark on T2 and T1

  - Treatment previously included medications, PT, and palliative ESIs. There has been growing evidence for the potential use of basivertebral nerve ablation. 5-year durability data from the SMART trial demonstrated visual analogue scale (VAS) reduction by 4.38 at 5 years and Oswestry disability index (ODI) reduction by 25.95, and one-third (34%) of patients were pain-free (Fischgrund et al., 2020). However, there is no consensus on the ideal therapy.

Spinal Stenosis

- Focal narrowing of the central spinal canal that causes compression of the spinal cord (cervical or thoracic) or spinal nerves and/or cauda equina (lumbar).
- The most common cause is degenerative changes (spondylotic ridging ventrally, facet hypertrophy laterally, ligamentum flavum hypertrophy posteriorly) that narrow the central spinal canal. Patients with lumbar spinal stenosis typically present with lower limb or buttock pain exacerbated by prolonged standing or walking and relieved with sitting and bending forward, including using a shopping cart.
- Other contributing factors: Superimposed disc herniation, spondylolisthesis, congenital stenosis, trauma, vertebral body fracture with retropulsion
- The degree of spinal stenosis does not necessarily correlate with the severity of a patient's symptoms.
- Clinical presentation: Axial spinal pain with or without radicular symptoms that worsens with standing/walking and improves with sitting or bending.
- Cervical stenosis should be routinely monitored clinically and radiographically to evaluate for progression and myelopathic changes.

🕮 Back Pain and Work

- There is a decreased likelihood of returning to work with increasing duration of back pain.
- Lifetime return to work rate for injured workers:
  - Off from work for 6 months: 50%
  - Off from work for 1 year: 25%
  - Off from work for 2 years: <5%

PAIN INTERVENTION

INTERVENTIONAL SPINAL PROCEDURES

Radiation Safety

- X-ray is a form of electromagnetic, ionizing radiation. The adverse effects of long-term radiation exposure include carcinogenesis, radiation burns, and cataract formation. These effects may take years to manifest.

- The biological effects of ionizing radiation are proportional to the time of exposure.
- Radiation exposure (Table 11–12) is inversely proportional to Uhe square of the distance from the radiation source.

TABLE 11–12 Radiation Exposure and Dose

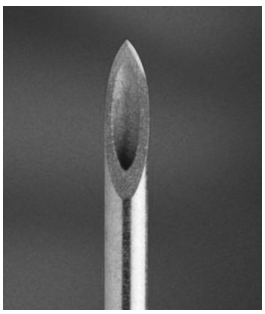
TERM	TRADITIONAL UNITS	SI UNITS	CONVERSION
Exposure	Roentgen	Coulomb/kg	1 R = 2.5 × 10^{-4} C/kg
Radiation absorbed dose	rad	Gray	100 rad = 1 Gray
Radiation equivalent in man	rem	Sievert	100 rem = 1 Sievert

SI, International System of Units.

- 🕮 Ways to minimize radiation exposure to the patient and practitioner:
  - Use of a lead apron, thyroid shield, and lead glasses is paramount to practitioner safety during fluoroscopy procedures. In addition, a film badge dosimeter worn outside of the lead protection should be utilized to monitor cumulative radiation exposure. The practitioner's extremities should also be kept as far away as possible during use of the fluoroscopy machine.
  - Use of a mobile lead shield and a lead table skirt can also help minimize exposure.
  - The x-ray tube should be positioned as far away from the patient as possible so a larger area is exposed to a smaller dose of radiation.
  - The x-ray image intensifier should be placed as close to the patient as possible to optimize image quality and minimize scatter radiation.
  - Collimation (narrowing of x-ray beam) reduces direct and scatter radiation exposure to the patient and practitioner.
  - Short Sulses of exposure (pulsed mode) rather than continuous exposure should be used when possible.
  - If fluoroscopy is used in manual mode, the kilovoltage peak (kVp) should be increased while minimizing the milliamps (mA).

Needle Selection

- Neuraxial procedures such as epidural injections of various approaches, facet MBBs, and some sympathetic blocks surrounding the spine will dictate selection of spinal needle length, gauge, and bevel/tip design.
  - All the needles described here are made with a tightly fitting, removable stylet. This ensures greater ease when directing the needle by preventing skin or tissue coring. This also reduces the chance of introducing epithelial tissue or contaminants at deeper target sites and prevents clogging of the needle lumen.
  - Once the needle is positioned, the stylet can be removed for injection of contrast or medication.
  - Available in a variety of sizes from 16 to 30 gauge. Most commonly used needles are 20 to 25 gauge. In general, higher gauge (smaller size) needles cause less patient discomfort and less risk of vascular injury but may require more skill to steer correctly.
  - Selection of appropriate needle length is based on patient habitus and angle of approach to desired injection target(s).
- **Quincke needle**: Lancet bevel tip with stylet and notched hub indicating bevel orientation cuts quickly through tissue; high degree of deflection allows for maneuverability around and into small spaces: Drives "away" from the notch.
  - Used for transforaminal (TF) epidural injections, MBBs, facet injections



Quincke needle tip

- **Tuohy needle**: Short bevel needle with a blunt, curved tip, winged hub, and centimeter depth markings, originally conceived for placement of epidural anesthesia catheters (Rand et al., 2016).
  - Curved short bevel tip minimizes inadvertent puncture of dura and provides tactile feedback important in driving needle through ligamentous structures of the interlaminar (IL) approach.
  - A winged hub allows for more force and directional control when driving the needle, and centimeter markings provide visual depth control.
  - Used for IL epidural injections

Tuohy needle tip

- **RFA cannula needle**: Insulated needle with tightly fitting stylet and exposed active tip designed to receive RFA electrode once placed at desired target
  - Manufactured in variety of gauge, lengths, active tip length, and tip design (sharp or blunt, straight or curved)
  - Lesion size is dictated by time, active tip length, and needle gauge (most significant variable), as well as duration of ablation (Benzon et al., 2011).

Medications

- Local anesthetics (Table 11–13):
  - Primarily function by reversibly blocking the sodium channels in nerve and muscle membranes
  - Signs and symptoms of systemic toxicity manifest as CNS and cardiac dysfunction.
  - **CNS toxicity**: Mild—lightheadedness/dizziness, perioral numbness, blurred vision, tinnitus, tremors, shivering; severe—tonic–clonic seizures, respiratory depression/arrest
  - **Cardiac toxicity**: Arrhythmias (conduction blocks, ventricular dysrhythmias), myocardial depression
  - When injected into the subarachnoid space, they have an effect on sympathetic nerves first (motor nerves last).
  - Intravascular anesthetic injection can cause adverse cardiovascular effects.

TABLE 11–13 📖 Local Anesthetics and Maximum Dosages

ANESTHETIC	DURATION OF ACTION	RECOMMENDED MAXIMUM DOSE
Bupivacaine (without Epi)	Long-acting 2–4 hours	2.5 mg/kg, not to exceed 175 mg
Lidocaine (without Epi)	Medium 30–60 minutes	4.5 mg/kg, not to exceed 300 mg
Ropivacaine	Long-acting 2–4 hours	5 mg, not to exceed 200 mg
Procaine	Short-acting 15–60 minutes	7 mg/kg, not to exceed 350–600 mg

- 📖 Corticosteroids (Table 11–14):
  - Cause direct inhibition of C-fiber neuronal membrane excitation and induce synthesis of a phospholipase A2 inhibitor, thereby preventing release of substrate for prostaglandin synthesis. This helps to reduce the inflammatory component of a radicular pain syndrome.
  - "May" also decrease back pain due to inflammation and sensitization of nerve fibers in the posterior longitudinal ligament and annulus fibrosus
- 📖 Nonparticulate steroids such United States dexamethasone are recommended over particulate steroids as first-line corticosteroids for epidural injections.
  - Dexamethasone, methylprednisolone, triamcinolone, and betamethasone have little or no mineralocorticoid activity. The increased glucocorticoid activity of dexamethasone and betamethasone can theoretically result in greater increases in blood glucose compared with methylprednisolone and triamcinolone.
- 📖 Agents used for neurolysis:
  - Alcohol: Concentration 100%, **hypobaric** compared with cerebrospinal fluid (CSF)
    - Positioning for subarachnoid neurolysis: Lay on unaffected side, semiprone.

TABLE 11–14 Steroid Preparations Available for Commercial Use in the United States

DRUG	BRAND NAME (HOW SUPPLIED)	[a]POTENCY (MG)	EQUIVALENT DOSE (MG)	HALF-LIFE (HOURS)
PARTICULATE STEROID PREPARATIONS				
Betamethasone sodium phosphate Betamethasone acetate suspension	Celestone Soluspan (Betamethasone sodium phosphate or acetate, 3 or 6 mg/mL)	25–30	0.6	35–54
Methylprednisolone	Depo-Medrol (20, 40, or 80 mg/mL)	5	4	12–36
Triamcinolone acetonide	Kenalog (10 or 40 mg/mL)	5	4	1.5–2
NONPARTICULATE STEROID PREPARATIONS				
Dexamethasone sodium phosphate	Decadron (4 or 10 mg/mL)	25–40	0.75	36–72

[a]Relative glucocorticoid potency compared with hydrocortisone (cortisol). All have minimal mineralocorticoid activity
Source: From Datta R, Upadhyay KK. A randomized clinical trial of three different steroid agents for treatment of low backache through the caudal route. *Med J Armed Forces India*. 2011;67(1):25–33. doi: 10.1016/S0377-1237(11)80007-9; Lamer TJ, Dickson RR, Gazelka HM, et al. Serum triamcinolone levels following cervical interlaminar epidural injection. *Pain Res Manag*. 2018:8474127. doi: 10.1155/2018/8474127; https://www.pdr.net/drugsummary/.

  - Phenol: Concentration 3% to 12% (usually 6%), **hyperbaric** compared with CSF
    - Positioning for subarachnoid neurolysis: Lay on affected side, semisupine.

Potential Spinal Injection Complications

- Common adverse effects: Injection site pain, temporarily increased radicular pain and paresthesias without morbidity, acute anxiety, lightheadedness, diaphoresis, flushing, nausea, hypotension, and nerve injury. Less common potential events could include vasovagal syncope and dural puncture.
- Infection: 1% to 2% of spinal injections
- Cardiovascular: Bleeding and dysrhythmias, epidural hematoma
- Neurologic:
  - Trauma to the spinal cord or nerve roots is a rare but serious complication.
  - Embolic infarction of the spinal cord and brain is theorized with intravascular injection. Of note, there have been case reports of neurologic injury and paralysis with the use of both particulate and nonparticulate steroid medications (Gharibo et al., 2016).
  - The Food and Drug Administration (FDA) issued a black box warning requirement that all injectable glucocorticoid product labels carry a warning stating that "serious neurologic events, some resulting in death, have been reported with epidural injection of corticosteroids," and "the warning did not distinguish any difference in the risk associated with the various injection approaches (interlaminar, transforaminal, and caudal), locations of spinal injection (cervical, thoracic, lumbar, and sacral), or glucocorticoid formulations (solutions and suspensions), because the data suggested that each approach, location, and formulation was associated with some risk of neurologic injury" (Racoosin et al., 2015).
- Respiratory: Oversedation, spinal anesthesia
- Urologic: Urinary retention
- Dural puncture: <0.5%. A headache due to the dural puncture presents as a positional headache that is worse when upright and improved when supine, is based on a clinical diagnosis.
- Treatment of acute adverse reactions:
  - Urticaria: Benadryl or Vistaril, cimetidine, or ranitidine. Consider preprocedure antihistamine and oral steroids in patients who have **nonanaphylactic** allergic reactions to iodinated contrast media.

- Facial and laryngeal edema: Epinephrine, IV fluids for hypotension, advanced cardiac life support (ACLS) protocol
- Bronchospasm: O_2, beta-agonist inhalers, epinephrine, IV fluids for hypotension, ACLS protocol
- Hypotension with tachycardia: Reverse Trendelenburg, IV fluids, epinephrine
- Hypotension with bradycardia: Reverse Trendelenburg, IV fluids, atropine
- Severe hypertension: Nitroglycerin, give phentolamine in pheochromocytoma
- Seizures: O_2, secure airway and oxygenation, diazepam or midazolam, phenytoin, ACLS protocol
- Pulmonary edema: O_2, diuretics, consider IV morphine use, transfer to ICU
- Anaphylactic reactions occur most often within 2 to 6 hours after an epidural injection. ACLS protocol should be implemented immediately. Any medication that has been known to result in anaphylaxis SHOULD NOT be given.
- 🕮 Local anesthetic toxicity symptoms:
  - Early: Perioral and tongue paresthesias, dizziness, orthostasis
  - Late: Muscle twitching, drowsiness, CNS depression, respiratory depression, tonic–clonic seizures, bradycardia, hypotension, cardiac arrhythmia (conduction block), cardiac arrest
- 🕮 Local anesthetic toxicity management: ACLS protocols, cardiac arrhythmia treatment, lipid emulsion therapy
- Corticosteroid side effects: HAs, insomnia, facial erythema, rash and pruritus, dizziness, low-grade fever, transient hyperglycemia, and hypertension

Epidural Steroid Injections

- One of the common interventional procedures is performed for treatment of an inflamed spinal nerve root causing radicular symptoms.
- 🕮 Dexamethasone, a nonparticulate steroid, has been recommended to be used as a first-line option for transforaminal ESIs given its safety profile and noninferiority compared with particulate steroids (Rathmell et al., 2015).
- The most common indication for ESIs is cervical or lumbar radicular symptoms corroborated by positive diagnostic studies that corroborate the claimant's symptoms and exam findings, including a specific radiographic correlate on MRI/CT and/or EMG confirmation of a radiculopathy. A trial of more conservative care should generally be performed first prior to consideration of a more interventional approach.
- As with surgical intervention, the success rate for ESIs decreases when performed for axial spinal pain alone. Such injections should be considered after a diagnostic nerve block to confirm the source ofithe axial pain generator.

EPIDURAL INJECTION APPROACHES

- Transforaminal Approaches (**Figures 11–4 to 11–6**):

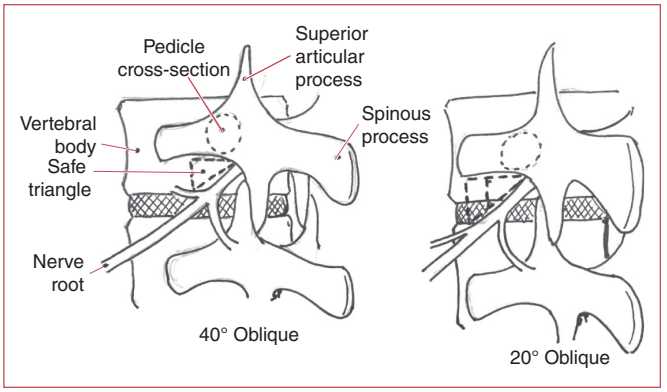


FIGURE 11–4 "Safe triangle" approach.

- Places medication near the anterior epidural space and closer to the inflamed spinal nerve root
- 🕮 The infrapedicular or **"safe triangle" approach**, in which the target endpoint for the spinal needle is inferior to the 6-o'clock position of the pedicle in the upper third of the neuroforamen in lumbar TF ESIs, has been traditionally advocated to avoid neural and vascular injury. Multiple views (anteroposterior [AP], lateral, oblique) are used to ensure proper spatial positioning.
- Placement of needle in epidural space is confirmed with epidural contrast flow under live fluoroscopy using multiple views (AP and lateral; **Figure 11–5**, AP view).

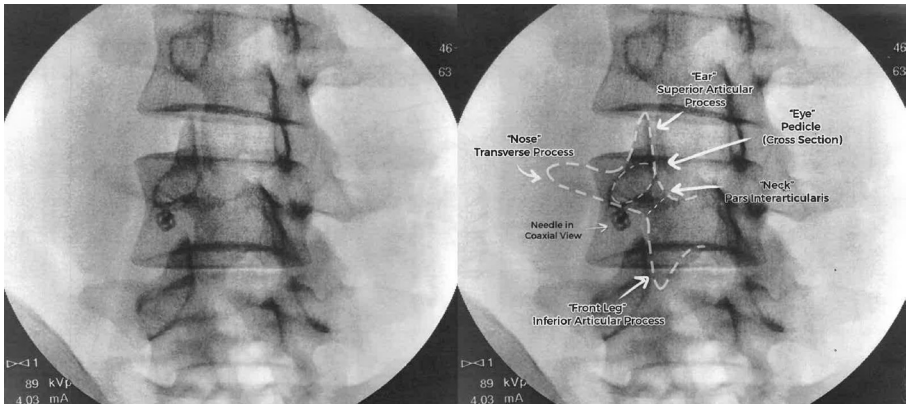


FIGURE 11–5 "Scotty dog" on fluoroscopy.

- 🕮 The borders of the safe triangle include:
  - Superior border: Horizontal line parallel to the inferior border of the pedicle
  - Lateral border: Lateral edge of the vertebral body
  - Hypotenuse: Spinal nerve root
- Newer literature has shown that spinal radicular arteries are most likely to be found in the upper and middle thirds of the foramen. Some studies instead suggest the use of Kambin's triangle, an infraneural approach to avoid intravascular injection.
- Cervical TF ESIs can be technically demanding and potentially hazardous for several anatomic reasons, including close proximity to the vertebral artery and penetrating radicular arteries relative to the spinal nerve. Extra safety measures are advocated during such procedures, including use of a nonparticulate steroid, digital subtraction angiography (DSA), and administration of a test dose anesthetic prior to injection of the corticosteroid.

• Interlaminar approach (**Figure 11–6**):

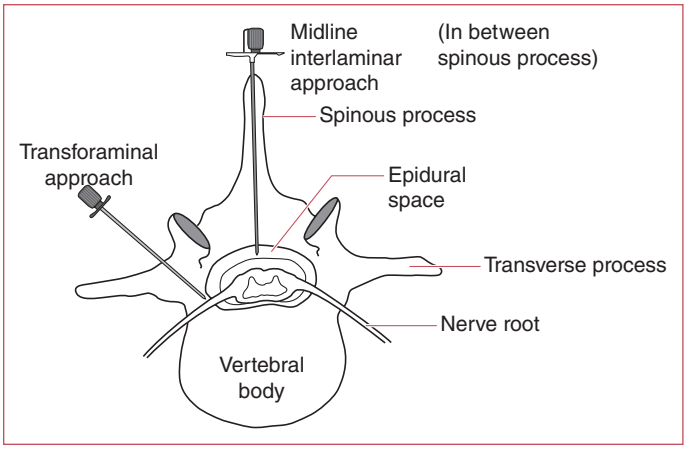


FIGURE 11–6 Transforaminal and Interlaminar approaches to epidural injections.

  - A midline or paramedian approach when the patient is placed in a prone position
  - Using an AP view, an 18- or 20-gauge Tuohy or Crawford needle connected to a loss of resistance (LOR) syringe is advanced into the IL space. A midline IL approach will advance the needle through the supraspinous ligament → interspinous ligament → ligamentum flavum → posterior epidural space.
  - Confirm placement of the needle in the epidural space with LOR in the LOR syringe and with proper epidural contrast flow under live fluoroscopy using multiple views (AP, lateral, contralateral oblique).
- Caudal approach:
  - 🕮 After the sacral hiatus is identified on palpation (medial to the sacral cornu) under fluoroscopy, the needle is advanced through the sacral hiatus. The needle should not be advanced higher than the S3 level, as the dural sac typically ends at the S2 level.
  - High injectate volumes are used to treat pathologic lesions superiorly up to the L4 to L5 or L5 to S1 levels. As a result, its efficacy is limited.
  - Has the least risk for dural puncture, but the medication is diffused to the greatest degree.

Diagnostic Medial Branch Blocks

- The medical literature notes the lack of clinical accuracy with diagnostic facet-mediated pain based on clinical history, physical exam maneuvers, and imaging findings (Dreyer & Dreyfuss, 1996).
- Diagnostic MBBs are diagnostic injections that anesthetize the medial branch nerves innervating a particular facet joint to confirm or rule out a diagnosis of facet-mediated pain syndrome.
- The primary pain generator in a confirmed facet syndrome diagnosis is the facet joint itself rather than the medial branches of the dorsal rami. As a result, "therapeutic" medial branch injections with corticosteroids would not be indicated even after a positive diagnostic block, as the diagnosis is not a medial branch neuropathy.
- A positive response is considered when there is >80% pain relief postprocedure. Performing two (an initial and a confirmatory) diagnostic MBBs is recommended, as studies have shown a high false-positive rate of >50% when only a single diagnostic block is performed and decreased efficacy of RFA procedures.
- **Cervical MBB:**
  - With the patient lying prone, the C-arm is oriented caudally without oblique angulation to visualize the facet joints and articular pillars. The needle is aimed at the articular pillars of the vertebra.
  - Use contralateral oblique view to confirm needle is not in the foramen.
  - The T1 transverse process (TP) articulates with the first rib and can be used to help identify the appropriate cervical level.
- **Lumbar MBB:**
  - With the patient lying prone, the C-arm is angled with slight oblique to optimally visualize the junction of the SAP and TP.
  - After the needle is positioned in the groove between the SAP and TP (where the medial branch lies) and placement is confirmed with contrast, a small amount (0.3–0.5 mL) of test anesthetic is injected. Pre- and postprocedure pain scores should be obtained to quantify the degree of pain relief.

Radiofrequency Ablation

- For patients with positive diagnostic MBBs, RFA can provide longer lasting pain relief than facet injections by denervating the medial branch nerves through thermocoagulation. In contrast, pulsed RF is a technique that uses lower temperatures to result in cellular changes but not permanent nerve tissue damage.

- **RFA (neurotomy):**
  - Treatment produces a focal area of thermal lesioning perpendicularly to the shaft of the active tip of an insulated cannula (Q-tip shape burn volume). Little destruction occurs at the end of the needle tip.
  - As a result, the active tip should be placed *parallel* to the target medial branch nerve to produce optimal nerve lesioning.
  - Performing RFA after two positive diagnostic MBBs results in significantly higher success rates, as there is a 50% false-positive rate with only one positive diagnostic MBB. It is not recommended to perform an RFA without diagnostic confirmation of the pain generator first given that clinical exam maneuvers and diagnostic imaging studies cannot accurately corroborate a facet syndrome.
  - Complications: Spinal nerve root lesioning (causing paralysis, neuropathic pain), increased pain, vasovagal reaction, and bruising. Ataxia (lesion to third occipital nerve), neuritis, and dropped head syndrome can occur in cervical RF. Other risks include infection and bleeding.
- **Pulsed RF:**
  - Pulsed RF applies intermittent RF energy to the tip of the cannula at much lower temperatures compared with RF ablation. It creates cellular alterations in the tissue without permanent damage.
  - The clinical efficacy of pulsed RF continues to be investigated.

Sacroiliac Joint Injections

- The SI joint is a diarthrodial joint with a complex and variable innervation.
  - Some studies suggest that the posterior SI joint receives its innervation from the L4 to S3 dorsal rami, while the anterior SI joint receives its innervation from the L2 to S2 ventral rami.
- The prevalence of SI joint-mediated pain in those with suspected SI joint pain diagnosed by fluoroscopically guided, contrast-enhanced diagnostic blocks is approximately 15% (Maigne et al., 1996; Schwarzer et al., 1995).
- Diagnostic intra-articular injections of local anesthetic into the SI joint can help to determine or rule out SI joint pain. Therapeutic injections should be considered only after the diagnosis is properly confirmed. Similar to facet syndrome, clinical exam and imaging findings are unreliable in diagnosing SI joint dysfunction. It does not diagnose SI joint complex pain, for which diagnostic lateral branch blocks would be indicated.
- Fluoroscopy is used to identify the inferior border of the joint at a point where the anterior and posterior joint lines overlap. Once the spinal needle is advanced to the inferior border of the joint, needle placement is confirmed with radiopaque contrast.

Lumbar Discography

- Discography is a diagnostic procedure in which a spinal disc is pressurized to establish or rule out a diagnosis of discogenic pain. Its diagnostic utility remains controversial given the high rates of false positives as well as potential long-term sequela of increased risk of accelerated disc degeneration.
- It is typically performed in patients presenting with axial spinal pain with or without radicular symptoms unresponsive to more conservative treatments, and diagnostic studies do not show definitive other corroborating pathology that correlates to the etiology of the pain.
- Procedure technique:
  - On an oblique fluoroscopic view with the SAP at the midline of the vertebral endplate, the needle is directed just anterior to the SAP of the inferior vertebral body level and advanced slightly into the disc. Note: A double needle technique can be used to minimize the potential for infection.
  - Needle placement in the center of the disc should be confirmed in multiple views (AP, oblique, and lateral).

  - A contrast agent should then be *slowly* injected to pressurize the disc. Pain response (intensity, location, and character) and pressure manometry levels are recorded. A control level should be performed.
- Procedure risks include infection (diskitis, osteomyelitis, abscess), accelerated disc degeneration, high false-positive results, and increased pain. Preprocedure IV and intradiscal antibiotics are recommended to prevent diskitis/osteomyelitis.
- **Interpretation:**
  1. ***Pain response:*** Most important information to gather is the patient's pain response to the procedure.
     - Pain level: Quantified measure of pain (such as VAS) pre- and postprovocation at that level
     - Character: Concordant versus nonconcordant
     - Location: Concordant location versus different location of pain
  2. ***Disc manometry:***
     - Disc manometry helps to quantify and standardize disc stimulation intensity.
     - Normal opening disc pressure: 5 to 25 psi
     - Above 30 psi usually indicates that the needle tip is in AF.
     - Excessive pressure (>50 psi above opening pressure) can result in false-positive results.
     - Can also characterize disc annulus sensitivity according to manometry.
  3. ***Disc annulus sensitivity:***
     - Chemical discs: Concordant pain response at 15 psi above opening pressure
     - Mechanical discs: Concordant pain response at 15 to 50 psi above opening pressure
     - Indeterminate discs: Pain provocation at 51 to 90 psi above opening pressure; should not be considered clinically significant
     - Normal discs: No pain provocation

Sympathetic Blocks

🕮 SYMPATHETIC NERVOUS SYSTEM

- Part of the autonomic nervous system designed to maintain the physiologic homeostasis of the body
- Presynaptic sympathetic fibers arise from the intermediolateral cell column horn of the spinal cord from T1 to L2 levels and leave the CNS via the ventral roots.
- The presynaptic sympathetic fibers can travel through to the white rami communicans and
  - Synapse at that level with paired sympathetic (**para**vertebral) ganglia (3 cervical, 11 thoracic, 4 lumbar, 4–5 sacral, and 1 fused coccygeal) in the sympathetic chain (**Figure 11–7**).
  - Travel cephalad or caudal levels to synapse with other paravertebral ganglia.
  - Pass through the sympathetic chain (without synapsing) as splanchnic nerves that synapse with **pre**vertebral ganglia (celiac, superior and inferior mesenteric, adrenal medulla ganglia).
    - Plexus network of postsynaptic fibers innervate abdominal and pelvic viscera (hypogastric, splanchnic, and mesenteric plexuses).
  - Exit with a spinal nerve via gray rami communicantes for peripheral distribution.
- Postganglionic axons are unmyelinated and leave the paravertebral ganglia via the gray rami communicantes and exit via the segmental spinal nerves.

🕮 STELLATE GANGLION BLOCKS

- The stellate ganglion comprises the lower cervical sympathetic and upper thoracic ganglia and is located anterolateral to the C7 vertebral body.
- It receives parasympathetic input and transmits sympathetic output to the head, neck, heart, and upper extremities.
- 🕮 Injection target area and clinical landmark is the anterior tubercle of the C6 vertebral body transverse process known as Chassaignac tubercle (aka carotid tubercle).
- Anomalous intrathoracic pathways known as *Kuntz's nerves* can bypass the stellate ganglion, resulting in incomplete sympathetic blockade.

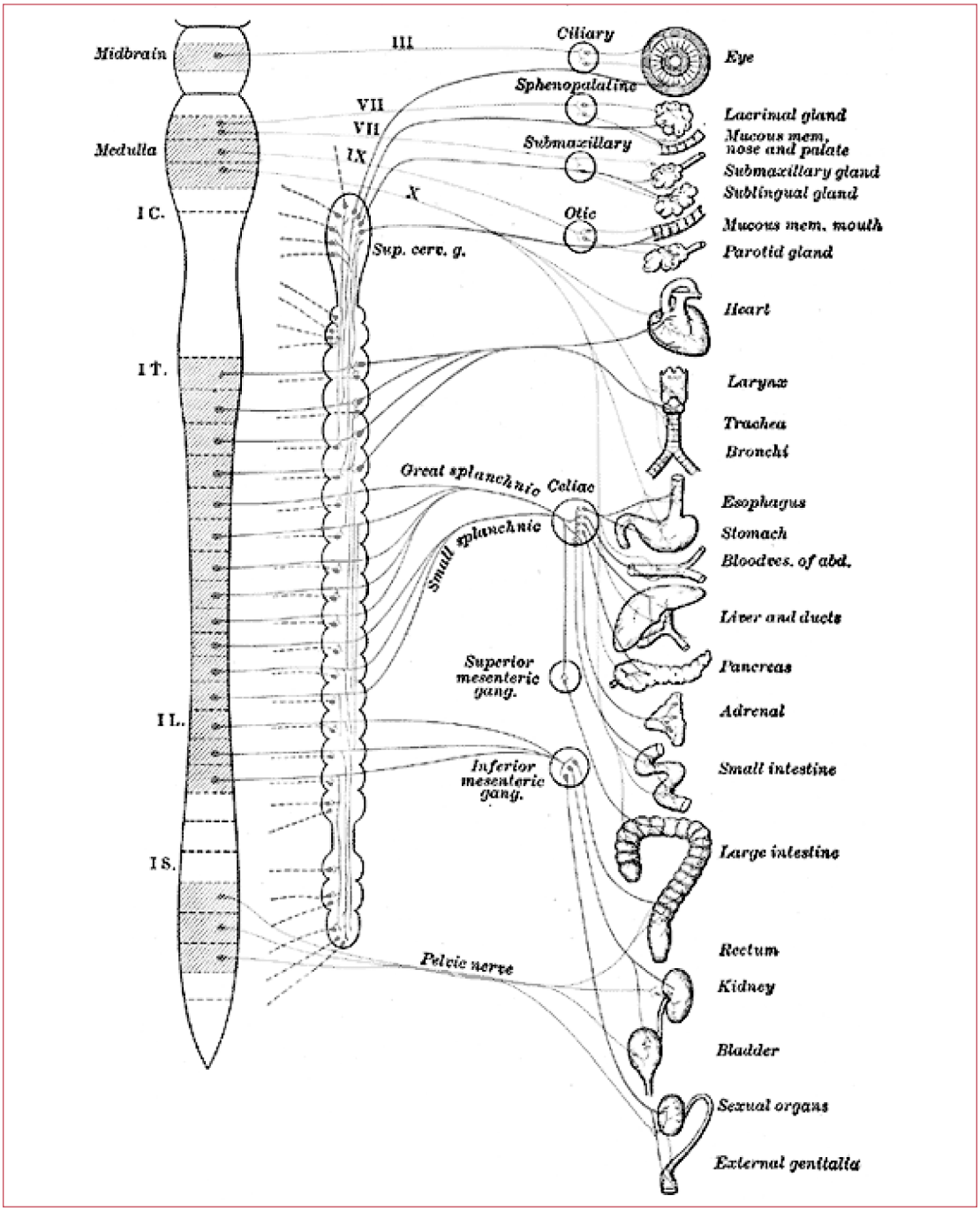


FIGURE 11–7 Sympathetic nervous system.
Source: From Gray H, Lewis WH, eds. *Anatomy of the Human Body*. Lea & Febiger; 2018.

- *Indications:*
  - Chronic facial and/or cervicobrachial pain syndromes, CRPS Type I or II, vascular and sympathetically mediated HAs (*Note: Stellate ganglion blocks can potentially exacerbate a migraine HA in certain situations), vascular insufficiency/vaso-occlusive diseases (e.g., Raynaud's syndrome), neuropathic pain syndrome, postherpetic neuralgia, trigeminal neuralgia, neuropathic orofacial pain, phantom limb pain, or hyperhidrosis

- ***Complications*** (even with a successful injection) can include:
  - 🕮 Ipsilateral Horner's syndrome (ptosis, miosis, anhidrosis, enophthalmos).
  - Hoarseness (local anesthetic spread to recurrent laryngeal nerve leading to vocal cord paralysis).
  - Paralysis: From brachial plexus block and epidural/subdural/intrathecal, or intravascular injection.
  - Dyspnea (local anesthetic spread to phrenic nerve leading to hemidiaphragm).
  - Intravascular/intrathecal injection of local anesthetic potentially leading to serious complications such as seizures and cardiac arrhythmias.
  - Organ injury, such as pneumothorax, esophageal perforation, vascular injuries (carotid, vertebral arteries).
  - Infection: Meningitis/epidural infection, cellulitis.
  - Hematoma formation.

THORACIC SYMPATHETIC BLOCKS

- Ten (sometimes 11) pairs of thoracic sympathetic ganglia are located paravertebral to the thoracic vertebral bodies.
- Cardiac plexus: T1 to T4 innervation
- Celiac ganglion: T5 to T12 innervation
- Aortic plexus: Comprises the aorticorenal ganglion as well as the superior and inferior mesenteric ganglia; it provides sympathetic innervation to various lower abdominal viscera

LUMBAR SYMPATHETIC BLOCKS

- Sympathetically mediated pain in the lower abdominal/pelvic regions or the lower extremities can be mediated by sympathetic blockade.
- ***Indications:***
  - Lower abdominal/pelvic genitourinary pain syndromes
  - Lower extremity pain (PVD, CRPS Type I or II)
  - Hyperhidrosis
- ***Complications:***
  - Vasodilation, increased temperature, reduction of edema
  - Genitofemoral neuralgia with chemical neurolysis > neuralgia with RF lesioning
  - Intravascular or subarachnoid injection

GANGLION IMPAR BLOCK

- Sympathetically mediated pain in the perineal region can be mediated by anesthetic blockade.
- Injection Technique: Insertion of Quincke spinal needle through the sacrococcygeal junction to a location anterior to this junction. Contrast is initially injected to confirm placement before an anesthetic or neurolytic agent is injected.
- ***Indications:*** Perineal or rectal pain, intractable in nature, and often cancer-related
- ***Complications:*** Possible neuritis, deafferentation pain, sensory changes, sexual dysfunction, and incontinence (rare)

Regional Blocks

CELIAC PLEXUS BLOCKS

- 🕮 Often indicated for controlling pain arising from the epigastric viscera, in particular from upper abdominal neoplasms such as pancreatic cancer.
- Receive sympathetic fibers from the greater, lesser, and least splanchnic nerves.
- Receive parasympathetic fibers from the vagus nerve.
- ***Indications:***
  - Sympathetically mediated thoracic, chest wall, upper abdominal viscera pain; hyperhidrosis; intractable cardiac arrhythmia; Prinzmetal's angina; acute (occlusion) or chronic vascular (Raynaud's) diseases; upper extremity CRPS; postthoracotomy pain; acute herpes zoster; postherpetic neuralgia; postmastectomy phantom breast pain

- *Complications:*
  - Pneumothorax (postprocedure chest x-ray is recommended), infection, or temporary intercostal neuritis

SUPERIOR HYPOGASTRIC PLEXUS BLOCK

- Treatment for chronic pelvic pain from gynecologic, colorectal, or genitourinary cancer
- 🕮 The hypogastric plexus is located at the lower one-third of the L5 vertebral body from the anterolateral border of L5 bilaterally.
  - L4 and L5 spinous processes are identified and needle insertion sites are 5 to 7 cm lateral to the midline at the level of L4 to L5 interspace.
  - With the bevel of the needle directed medially, two 7- to 9-in., 22-gauge needles are inserted under fluoroscopic guidance so that the needle tips lie anterolateral to the L5 to S1 intervertebral space.
  - In the AP view, the spread of contrast should be confined to the midline region.
  - In the lateral view, a smooth posterior contour corresponding to the anterior psoas fascia indicates needle is at the proper depth.
  - Diagnostic block with anesthetics and neurolytic block with phenol or alcohol

SACRAL NERVE BLOCKS

- Treatment for visceral pelvic pain. Sacral nerves provide sensory innervation to the anorectal region and motor innervation to the external anal sphincter and levator ani muscles. S1 to S4 provide the majority of visceral innervation to the bladder, urethra, and external genitalia.

Neuromodulation

SPINAL CORD STIMULATION

- 🕮 The Gate Control Theory of Pain by Ron Melzack and Patrick Wall explains the experience of pain pathophysiologically as a complex interplay of the PNS with the CNS. Pain messages are modulated by "nerve gates" along the nerve pathways. SCS utilizes the Gate Control Theory by introducing external neuromodulation in the form of nonpainful electrical signals from percutaneous electrodes placed over the dorsal columns.
- Stimulation increases with an increase in amplitude, frequency, pulse width, and supine position.
- Nerve root stimulation increases incrementally in longitudinal stimulation and axial stimulation increases incrementally in transverse stimulation.
  - Common indications: Failed back surgery syndrome (FBSS), discogenic pain refractory to conservative and surgical interventions, CRPS, arachnoiditis, painful peripheral neuropathy (including diabetic peripheral neuropathy), refractory angina pectoris, nonoperable ischemic limb pain, and migraine HA
- Electrode insertion levels:
  - 🕮 Should be at least two spine segments below the target level so that at least 3 in. of the lead body lies within the epidural space to maximize electrode stability.
  - Cervical electrode placement: Below T1 to T2 vertebral body level
  - Lower part of the body: T12 to L1 or L1 to L2
  - Upper extremity: T2 to T3 or T3 to T4
- **Intraoperative testing: It is important that the patient is awake and cooperative to optimize sensory and motor testing.**

DORSAL ROOT GANGLION STIMULATION

- Selected DRGs are electrically stimulated. The mechanism is similar to SCS, which is based on the Gate Control Theory.
- Advantages of DRG stimulation include selective and focused targeting of affected dermatomal distributions. It also results in predominantly a nonparesthesia-based therapy compared with traditional SCS.

- Common indications: CRPS, neuropathic pain conditions affecting specific regions of the body lasting 6 months or longer (e.g., isolated foot or groin pain)
- ACCURATE Trial in 2017 showed that DRG provided a higher rate of treatment success for CRPS compared with SCS. In addition, DRG also resulted in more targeted therapy to painful parts of the lower extremities with less postural variability in paresthesia intensity compared with SCS.
- Similar to SCS, DRG stimulation begins with a trial lasting 5 to 7 days. A successful trial may be followed by permanent implantation of the DRG stimulation device.

Implanted Drug Delivery Device

- Patients with chronic intractable pain with known pathophysiology who failed maximal medical and/or surgical therapy may be considered as candidates for an implanted drug delivery system device.
- Pain must be sensitive to the infused agent(s) and patient must have had a favorable response to a trial of intraspinal administration of the agent(s).
- Patient should also have a favorable psychosocial evaluation and have a life expectancy of >3 months.
- Intraspinal administered drugs used:
  - Opioids (e.g., morphine, hydromorphone, fentanyl)
  - Alpha-adrenergic agonists (e.g., clonidine and tizanidine)
  - GABA-B agonists (e.g., baclofen)
  - Local anesthetics (e.g., bupivacaine and ropivacaine)
  - NMDA agonists (e.g., ketamine)
  - Others (e.g., ziconotide)
- Routes of administration: Intrathecal versus epidural
  - Intrathecal has increased risks of HA, neural injury, and supraspinal spread, but requires lower dosage, less systemic effect, and no catheter tip dural fibrosis.
- 🕮 Complications:
  - Infection
  - Skin erosion from hardware
  - Pump failure
  - Catheter kinking, migration, and obstruction
  - Catheter tip granuloma formation (inflammatory process)

🕮 VISCOSUPPLEMENTATION (HYALURONIC ACID) INJECTIONS

- Hyaluronic acid is a naturally occurring gel-like substance that acts as a lubricant as well as a shock absorber in joints. Intra-articular hyaluronic acid injections have been U.S. FDA approved only for the treatment of symptomatic knee OA.
- Many hyaluronic acid formulations are derived from rooster combs (**Note: Older, nonsynthetic hyaluronic acid formulations are contraindicated in patients with egg or chicken allergies). Newer formulations are synthetically derived and do not have the same allergy concerns.
- Despite FDA approval for treatment of knee OA, more recent studies have varied with respect to statistically significant outcomes and clinically relevant efficacy.

OTHER PERIPHERAL NERVE BLOCKS

Suprascapular Nerve Block

- Indicated for the relief of acute and chronic shoulder pain secondary to arthritis, capsular tear, or adhesive capsulitis
- The suprascapular nerve originates from brachial plexus (primarily C5–C6, sometimes C4) and can be best accessed as it travels over the suprascapular notch before diving into the supraspinatus and infraspinatus. The nerve supplies sensory innervation to approximately 70% of the joint.

- Technique:
  - Ultrasound guidance can be utilized to best visualize the suprascapular nerve traveling within the notch. Ultrasound is less expensive and free of radiation exposure when compared with CT guidance or fluoroscopy and allows for live visualization of the nerve.
  - Needle entry is achieved by locating the midpoint of the scapular spine and drawing a vertical line bisecting it, which approximates the location of the scapular notch. Needle entry is lateral to this point directed medially and anteriorly.
  - Pneumothorax is a rare complication.
  - Care must be taken to not injure the suprascapular artery.

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ASSOCIATED TOPICS IN PHYSICAL MEDICINE AND REHABILITATION

12

SPASTICITY—Elie Elovic, MD • Edgardo Baerga, MD • Steven V. Escaldi, DO • Kristen A. Harris, MD

MOVEMENT DISORDERS—Elie Elovic, MD • Edgardo Baerga, MD • Roger Rossi, DO • Steven Markos, MD • Jessie Chan, MD

WHEELCHAIRS—Steven Kirshblum, MD • Mary T. Shea, MA, OTR, ATP • Beverly Hon, MD

OSTEOPOROSIS—Barbara Hoffer, DO • Sara J. Cuccurullo, MD • Krishna J. Urs, MD • Casey Schoenlank, MD • Eric Heckelmann, MD

BURNS—Alan W. Young, DO • Kristen A. Harris, MD • Shirley Shen, MD

BIOSTATISTICS—Joseph Lee, MD • Kathy Kalmar, PhD • Bart K. Holland, PhD • Heather Platt, MD • Hayk Petrosyan, PhD

ETHICS—Lyn Weiss, MD

MULTIPLE SCLEROSIS—David S. Rosenblum, MD

ULTRASOUND—Steven V. Escaldi, DO • Craig Van Dien, MD • Sagar Parikh, MD

SPASTICITY

- **Spasticity** can result from numerous conditions, such as stroke, cerebral palsy (CP), anoxic brain injury, traumatic brain injury (TBI), spinal cord injury (SCI), multiple sclerosis (MS), and other central nervous system (CNS) neurodegenerative diseases.
- 🕮 **Spasticity** is a motor disorder characterized by an abnormal, *velocity-dependent* increase in the tonic stretch reflexes (muscle tone) with exaggerated phasic stretch reflexes (tendon jerks,

clonus) resulting from hyperexcitability of the stretch reflex. It is one component of the upper motor neuron syndrome (UMNS; Lance et al., 1980). This definition is best thought of as a classic neurophysiologic description of spasticity.

- Spasticity from cerebral origin results from an imbalance of descending inhibitory signals from the dorsal reticulospinal tract (RST) and the excitatory signals from the medial RST and vestibulospinal tract on Ia afferent fibers.
- The functional impairments seen in patients with spasticity result from the combined influences related to decreased intrinsic viscoelasticity of muscle, weakness, and motor overactivity.
- The definition of spasticity has broadened over the years to include factors that address both the neurophysiologic and clinical presentations. This evolution of the definition has resulted in a number of other terms that are used interchangeably with spasticity. Examples include "consequences of the UMNS" (Esquenazi, 2015), "disordered sensi-motor control" (Pandyan et al., 2005), and "involuntary muscle hyperactivity" (Dressler et al., 2018).

DEFINITIONS

- **UMNS:** A collective term that refers to different motor abnormalities produced in patients who have lesions proximal to the alpha motor neuron (spinal cord, brain), resulting in the loss of descending inhibition and hypersensitivity of the reflex arc in the spinal cord. It is characterized by a series of positive and negative signs described below.
- **Muscle tone:** The resistance of muscle to stretch or passive elongation
- **Hypertonia:** Subjective description of tone greater than normal
- **Associated reactions:** Abnormal contraction distant from the muscles involved in voluntary effort; contributes to synkinesia, overflow
- **Dystonia:** Movement disorder characterized by muscle contraction resulting in twisting, turning, and posturing
- **Spastic dystonia:** Muscle contraction present at rest, which leads to a persistent clinical posture that is sensitive to stretch. It describes a coexistence of dystonia and spastic features in the same muscle group.
- **Clonus:** Alternating muscle contraction and relaxation of agonist and antagonist muscles generated after rapid stretch.
- **Rigidity:** Resistance to stretch that is *not* velocity-dependent—the examiner feels the same resistance to stretch irrespective of the velocity at which a muscle group is being stretched.
- **Tremor:** A rhythmic, unintended movement of a body part. May be classified based upon context into four basic types: Rest, postural, action, and intentional (discussed further in the Movement Disorders section).
- **Myoclonus:** Sudden jerky motions that may be singular or repeated. Can be differentiated from tremor as myoclonus is less rhythmic and less predictable than a tremor (discussed further in the Movement Disorders section).
- **Spastic Co-contraction:** Abnormal antagonist contraction present during voluntary agonist effort, dependent on tonic stretch on antagonist
- **Associated reactions:** Involuntary contraction in a limb occurring distant from the muscles involved in voluntary effort. It occurs due to disinhibited spread of voluntary motor activity into a limb affected by a UMN lesion (i.e., synkinesis).
- **Released flexor reflex afferents:** Abnormal sensorimotor reflexes (Babinski's sign and flexor spasms)
- **GABA:** Gamma-aminobutyric acid (GABA), a neutral amino acid that acts as the main inhibitory neurotransmitter in the CNS. It is an agonist at two receptors: $GABA_A$ and $GABA_B$.
  - Activation of the $GABA_A$ receptors is inhibitory via an increase in chloride conductance.
  - There are two subtypes of $GABA_B$ receptors: The presynaptic receptors are inhibitory by mediating a decrease in Ca^+ conductance when activated; the postsynaptic receptors provide inhibitory input by increasing K^+ conductance when activated.

Manifestations of Upper Motor Neuron Syndrome

POSITIVE SIGNS (ABNORMAL BEHAVIORS)	NEGATIVE SIGNS (PERFORMANCE DEFICITS)
• Spasticity • Hyperreflexia • Clonus • Persistence or reappearance of primitive reflexes (e.g., positive Babinski response) • Rigidity • Spastic dystonia • Spastic co-contraction • Associated reactions • Increased cutaneous reflexes	• Muscle weakness • Paralysis/paresis • Loss of voluntary movement • Loss of coordination • Decreased dexterity • Muscle atrophy • Muscle fatigability

GRADING OF DEEP TENDON REFLEXES

GRADE	DESCRIPTION
0	Absent
1	Diminished but present; minimal
2	Normal
3	Brisk and excessive
4	Very brisk, often with rhythmic reflex contractions (clonus)

ASSESSING SPASTICITY AND TREATMENT EFFICACY

- There are two commonly used assessment scales in clinical practice. They are the Modified Ashworth Scale (MAS) and the Tardieu Scale.
- **MAS** is a widely used qualitative scale for the assessment of tone.
  - It measures the resistance to passive stretch **(Table 12–1),** but it does not account for difference in velocity or contracture.

TABLE 12–1 Modified Ashworth Scale for Spastic Hypertonia

0	No increase in tone
1	Slight increase in muscle tone, manifested by a catch and release or minimal resistance at the end of the ROM when the affected part(s) is moved in flexion and extension
1+	Slight increase in muscle tone, manifested by a catch, followed by minimal resistance throughout the remainder (less than half) of the ROM
2	More marked increase in muscle tone through most of the ROM, but affected part(s) easily moved
3	Considerable increase in muscle tone, passive movement difficult
4	Affected part(s) rigid in flexion or extension

ROM, range of motion.

- **Tardieu Scale:**
  - The Tardieu Scale assesses the muscle while completely relaxed, identifying the angle where passive stretch is arrested at a slower and faster velocity of stretch (see below). It may better assess spasticity and differentiate spasticity from contracture.
  - Evaluating muscle at different velocities helps distinguish musculotendinous stiffness from spasticity.
  - For each muscle group tested, the joint is moved at three different velocities (V1, V2, V3). The quality of muscle reaction and angle at which muscle reaction occurs are recorded.

  - Velocities:
    - V1—Joint is moved as slow as possible.
    - V2—Speed of limb segment falls with gravity.
    - V3—Joint is moved as fast as possible.
  - Muscle response quality:
    0—No resistance through range of motion (ROM)
    1—Slight resistance throughout ROM, no clear catch
    2—Clear catch interrupting passive movement, followed by release
    3—Fatigable clonus (<10 seconds)
    4—Nonfatigable clonus (>10 seconds)
    5—Joint is immovable
- **Modified Tardieu Scale:**
  - R1 and R2 components added to Tardieu Scale
  - R1 is defined as the angle at which muscle reaction occurs during V3 (fastest velocity).
  - R2 is defined as the angle at which muscle reaction occurs during V1 (slowest velocity).
  - A large difference between R1 and R2 indicates that velocity-dependent tone (i.e., spasticity) is predominant, while a small difference between R1 and R2 indicates that velocity-independent tone (such as from muscle contracture) is predominant.
- **Assessing spasticity treatment:**
  - It is critical that the metric selected to evaluate the efficacy of the spasticity treatment is relevant to the intervention and the specific goals of the treatment.
  - Potential assessment categories/parameters:
    - Goal Attainment Scale: A method of assessing the degree of achievement in reaching established rehabilitation goals
    - When the patient and family are involved in choosing the target goal, then the potential to address issues that are important to them can be achieved.
  - Physiologic parameter (e.g., H-reflex/M-response [H/M] ratio, muscle activity)
  - Passive activity (e.g., ROM, Ashworth, Tardieu)
  - Voluntary activity (e.g., Fugl-Meyer, box and block, Barthel index)
  - Functional measures (e.g., timed walking, Physiological Cost Index)
  - Quality of life measures (e.g., 36-Item Short Form Survey [SF-36], Satisfaction With Life Scale)

Complications Due to Spasticity (Katz, 1997)

- Interference with function/impedance of volitional movement
- Can cause extreme discomfort/pain in patients with intact (or abnormally increased) sensation
- Interferes with hygiene and nursing care
- Contractures and disfigurement
- Increased risk for development of decubitus ulcers
- May cause bone fractures; once present, malunion may occur due to forces
- Joint subluxation/dislocation
- Increased risk of heterotopic ossification (HO)
- Acquired peripheral/entrapment neuropathy

Benefits of Spasticity

- May help some patients with ambulation, standing, or transfers (e.g., stand pivot transfers)
- May assist in maintaining muscle bulk due to muscular contraction especially in muscles without voluntary motor control
- May assist in preventing deep vein thrombosis (DVT) and edema by providing improved venous flow secondary to muscle contractions
- May assist in preventing osteoporosis
- Decrease of pressure ulcer formation over bony prominences
- Can serve as a "diagnostic tool" for noxious stimuli (spasticity can be a sign of exposure to a noxious stimuli—infection, bowel impaction, urinary retention, etc.)

Treatment of Spasticity

CONSIDERATIONS

- As previously noted, spasticity may confer a functional benefit that must be taken into account prior to treatment. Additionally, the expectations and goals of all involved (patient, family, therapists, and physician) must be discussed prior to efforts to treat the patient's spasticity.
- As the treatment of spasticity is primarily aimed at ameliorating the positive symptoms and usually does not change the negative symptoms, all involved must be in agreement with the intended goals of treatment.
- The underlying pathology that is the cause of spasticity, time post-onset, overall medical status, support system, resources, and cognitive status all play some role in the treatment decision.
- The distribution of the spasticity, focal versus generalized, will affect the treatment decision.
- Treatment options represent a continuum of care (conservative ⟶ surgical):
  - Rehabilitation measures: Noxious stimuli avoidance, stretching/ROM, splinting, serial casting, modalities
  - Systemic medications
  - Local interventions (botulinum toxin/phenol/alcohol)
  - Surgical procedures
- Often, the best treatment strategy may require a nonserial or concomitant approach of the previously mentioned options.

PREVENTION

- The viscoelasticity of the muscle and tendon plays a role in spasticity so it is important to maintain daily stretching/ROM program.
- Avoid noxious stimuli such as infection, pain, DVT, HO, pressure ulcers, urinary retention or stones, and ingrown toenails.
- Education of the patient, their family, and any other caregivers, as well as other clinicians, is essential in the management process.
- Emphasize proper positioning, daily skin inspection, adequate bladder/bowel programs, and so forth.

MODALITIES: PHYSICAL MODALITIES AND THERAPEUTICS

- Stretching
- Splinting and serial casting
- Heat and cold modalities
- Proper positioning
- Direct tendon pressure
- Functional electrical stimulation
- Vibration
- Relaxation techniques
- Motor reeducation
- Biofeedback
- Extracorporeal shockwave therapy

PHARMACOTHERAPY IN SPASTICITY (TABLE 12–2)

- Currently, only four oral prescription medications are approved by the Food and Drug Administration (FDA) for the management of spasticity due to CNS disorder: Baclofen, tizanidine, dantrolene, and diazepam (see **Table 12–2**). Clonidine is also addressed in **Table 12–2**. Even though presently clonidine is not currently FDA-approved for treatment of spasticity, it is used in clinical practice. Other medications discussed later may also be effective.
- Systemic medications usually provide the most benefit in mild to moderate spasticity and are a better choice for generalized rather than localized spasticity.
- They may have their best effects in spasticity due to SCI or MS.
- They may reduce tone and decrease pain, but minimal evidence exists for improvement of function.

TABLE 12–2 Antispasticity Medications

MEDICATION	MECHANISM OF ACTION	SIDE EFFECTS	USES	CAUTIONS	DOSE
Baclofen (Lioresal)	• 🕮 Oral GABA-mimetic agent that acts as a GABA agonist at $GABA_B$ receptors • Provides inhibitory effects in the monosynaptic and, to a lesser extent, the polysynaptic reflex pathways • Activation of presynaptic $GABA_B$ receptors reduces Ca^+ influx and suppresses the release of excitatory neurotransmitters from the presynaptic axon • Activation of the postsynaptic $GABA_B$ receptors increases K^+ egress, maintaining membrane polarization. This makes it more difficult to depolarize the postsynaptic cell and helps to dampen the effects of any excitatory neurotransmitters released from the presynaptic axon. This reduced gamma motor neuron excitability reduces input to intrafusal muscle fibers and decreases muscle spindle sensitivity • May also reduce substance *P* in the spinal cord, contributing to decreased pain	Generally mild • 🕮 Sedation/drowsiness Patient may develop tolerance to this over time. May be dose limiting factor • Lowered seizure threshold • Weakness • GI symptoms • Tremor • Insomnia • Confusion	Drug of choice for: • Spinal forms of spasticity and MS • Also used in spasticity due to acquired brain injury, but no literature has demonstrated benefit • Studies suggest greater effect on LE tone than on upper extremity tone	• 🕮 Sudden withdrawal of drug can cause seizures, hallucinations, and rebound spasticity with fever • Renal clearance may require dose adjustment in renal disease • Requires slow taper to discontinue; otherwise, risk of seizure and hallucinations • Change from oral baclofen to intrathecal administration may be associated with withdrawal due to the relatively low cerebral concentration with intrathecal use as compared with oral • Theoretical interference with recovery with acquired brain injury	• Start 5 mg twice a day or three times a day; increase dose by 5 mg every 3–5 days up to 80 mg/day • 80 mg/day is the FDA's recommended max. dose; higher doses are frequently used and tolerated.
Diazepam (Valium)	• Does not substitute for GABA, but rather facilitates GABA's effects on the $GABA_A$ receptor subtype; overall effect results in membrane hyperpolarization and decreased neuronal firing • Acts diffusely throughout the neuroaxis. Net effect is increased presynaptic inhibition and reduced mono- and polysynaptic reflexes • Other benzodiazepines (clonazepam and clorazepate as a pro-drug) also used for spasticity management with similar mechanisms	• Sedation (most sedating of commonly used oral spasticity medications) • Memory impairment • Decreased REM sleep	• Demonstrated benefit in SCI and MS spasticity	• Usually unsuitable in TBI patients because of the adverse effects on attention and memory • Can have very long half-life due to active metabolites • Hepatic metabolism; clearance may be affected by concurrent use of other hepatically metabolized medications • CNS depression worsened with concurrent use of ETOH • Overdose can be treated with flumazenil • May have withdrawal symptoms if not tapered • Theoretical interference with recovery with acquired brain injury	• Start with a bedtime dose of 4 mg or 2 mg twice a day and titrate up, with a maximum dose of 60 mg/day • Fast-acting, allows for rapid titration to effective dose

TABLE 12–2 Antispasticity Medications (*continued*)

MEDICATION	MECHANISM OF ACTION	SIDE EFFECTS	USES	CAUTIONS	DOSE
Dantrolene sodium (Dantrium)	• 🕮 Unique among spasticity medications with peripheral site of action, acting at the level of the muscle itself • Acts peripherally in the striated muscle by blocking the release of Ca^{++} at the Ryanodine receptor in the sarcoplasmic reticulum • Reduces extrafusal muscle fiber contraction strength and muscle spindle sensitivity • Fast twitch motor units are more sensitive than slow twitch motor units • Minor effects on smooth or cardiac muscle	• 🕮 Liver toxicity in approximately 1% of patients. Highest risk in females over 30 years, on high doses (>300 mg), for >2 months. Risk of hepatonecrosis • Drowsiness/sedation (mild to moderate), less central effects given mechanism of action • Weakness, fatigue, paresthesias, diarrhea, nausea, vomiting	• Classically preferred drug for spasticity of cerebral origin (stroke, CP, head injury) • This may change as more experience with tizanidine • Use in SCI and MS may be limited by associated weakness • Also used to treat malignant hyperthermia, neuroleptic malignant syndrome, and hyperthermia due to baclofen withdrawal	• LFTs should be monitored (hepatotoxicity) • Potential interaction with nondihydropyridine calcium channel blockers	• Start 25 mg twice a day up to a total dose of 400 mg/day (100–200 mg/day usually appropriate). Usually dosed twice a day or three times a day
Clonidine (Catapres)	• Central acting alpha$_2$-adrenergic agonist with effects in locus ceruleus. In the spinal cord, provides presynaptic inhibition of sensory afferents • Commonly used as a transdermal patch; oral also available	• Hypotension/syncope • Nausea • Sedation • Depression • Dry mouth • Edema	• Largely studied in treatment of SCI-related spasticity • Reported useful in treatment of supraspinal (brainstem) spasticity • Other common uses include treatment of hypertension	• 🕮 Withdrawal following long-term, high-dose use can result in severe hypertensive crisis due to increased sympathetic activity • May impair hypoglycemia-induced tachycardia in patients with diabetes, thus delaying the diagnosis of clinically significant hypoglycemia in this population • Can theoretically impair poststroke motor recovery	• Transdermal patch: Start Catapres®—TSS with 0.1 mg patch every week up to 0.3 mg patch every week. Patches changed every 7 days • Oral: Start with 0.05 mg orally twice a day, may increase up to 0.4 mg/day

(*continued*)

TABLE 12–2 Antispasticity Medications (*continued*)

MEDICATION	MECHANISM OF ACTION	SIDE EFFECTS	USES	CAUTIONS	DOSE
Tizanidine (Zanaflex)	• Central acting alpha-2 adrenergic agonist, chemically related to clonidine • Binds to alpha-2 receptors both spinally and supraspinally • Thought to act by enhancing presynaptic inhibitory modulation of spinal reflexes. This effect is especially noted in polysynaptic reflexes • Less potent than clonidine in regard to blood pressure; less issues with hypotension	• Sedation/drowsiness, noted in up to 50% of patients • Potential for liver damage • Hypotension • Dry mouth • Bradycardia • Dizziness	• Spasticity due to SCI, MS, or acquired brain injury • Clinical trials show tizanidine to be as effective as oral baclofen or diazepam with a better overall tolerability • Fewer complaints of treatment-associated weakness than other spasticity medications	• LFT monitoring suggested (metabolized in liver) • Effective dose is patient-dependent and varies widely • Requires frequent dosing (short half-life) • Contraindicated for concurrent use with IV ciprofloxacin due to inhibition of cytochrome P450	• Start at 2–4 mg/day, usually at bedtime • May increase dose and frequency up to max 36 mg/day depending on patient tolerance

CNS, central nervous system; CP, cerebral palsy; ETOH, ethyl alcohol; FDA, Food and Drug Administration; GABA, gamma-aminobutyric acid; GI, gastrointestinal; IV, intravenous; LE, lower extremity; LFT, liver function test; MS, multiple sclerosis; REM, rapid eye movement; SCI, spinal cord injury; TBI, traumatic brain injury; TSS, toxic shock syndrome.

Other Oral Agents

The following is a partial list of the wide array of medications tried for the management of spasticity:

- Gabapentin (Neurontin®): A structural derivative of GABA with an uncertain mechanism of action used for management of neuropathic pain as well as spasticity. This may be a good choice as a second-line agent. Doses are commonly 300 to 900 mg three times a day.
- Antiepileptic medications: Topiramate (Topamax®), lamotrigine (Lamictal®), and phenytoin (Dilantin®)
- Cyproheptadine (Periactin®): Nonselective histamine and serotonin antagonist. Some trials have demonstrated comparable antispasticity effects to baclofen and clonidine. Primarily used in the SCI population.
- Cyclobenzaprine (Flexeril®): Acts primarily within the CNS at the level of the brainstem (as opposed to the spinal cord level), though its action in the latter may contribute to its overall skeletal muscle relaxant activity. Evidence suggests that the net effect of cyclobenzaprine is a reduction of tonic somatic motor activity, influencing both gamma and alpha motor systems.
  - In general, skeletal muscle relaxants are not indicated for the treatment of spasticity.
- Cannabinoids: D9-tetrahydrocannbinol and cannabidiol (CBD) are the active ingredients in marijuana. Evidence demonstrates that cannabinoids can improve spasticity by inhibiting the release of excitatory neurotransmitters, such as glutamate, and enhance the effects of GABA. May cause sedation, hypotension, and mood disorders including anxiety, psychosis, and impaired executive function.
- Morphine

LOCAL INTERVENTIONS

- Local interventions include *diagnostic nerve blocks, chemical neurolysis, chemodenervation* with botulinum toxin, and *motor point blocks*. These procedures are the best choice for management of focal/local spasticity issues or when the systemic effects of oral agents are prohibitive at required treatment doses.
- Local anesthetics and neurolytic agents can be applied to motor or mixed peripheral nerve branches, and neurotoxins can be applied to motor points to decrease spasticity.

Diagnostic Nerve Blocks

- Local anesthetics are injected perineurally, usually under the guidance of electrostimulation and/or ultrasound (US), to block nerve conduction for a period of hours.
- May be useful in planning for more permanent interventions such as chemoneurolysis, botulinum toxin, or even surgery. The temporary reduction in spasticity induced by these agents allows assessment of the potential benefit to be had from longer lasting procedures. It may also allow for evaluation of the functional utility of the spasticity.
- Local anesthetic agents work by blocking the voltage-gated sodium channels on the axon. This prevents depolarization of the axon membrane and interrupts transition of the signal along the axon.
- It is a useful means of assessing dynamic muscle resistance, but because it may also impair sensation, it may impact the ability to fully assess the benefits of spasticity reduction.
- Contraindications: Do not inject through infected or uncleanable skin.
- Anesthetic agents:
  - Lidocaine (Xylocaine®): An amide local anesthetic usually available in 1% or 2% concentrations and a length of action of 1 to 2 hours
  - Bupivacaine (Marcaine®): Longer acting amide local anesthetic; 0.25% or 0.5% concentrations commonly used. Length of action up to 10 hours.

Chemoneurolysis

- Chemical neurolytic agents can block spasticity for months to years. These agents induce demyelination and axonal destruction via protein denaturation and axonal necrosis.
- These agents may be used to disrupt nerves within a trunk (mixed nerve or motor nerve block) or at the motor point where it attaches to the muscle.

- Generally, injection of the agent utilizes electrostimulation and/or US guidance and requires technical skill to perform effectively.
- 🕮 Agents:
  - **Phenol:** Common concentrations used are from 2% to 7%. Lower concentrations (1%–3%) produce a transient effect due to demyelination with little axonolysis. For chemical denervation, higher concentrations (>5%) are required. The effects of this increased concentration are neurolytic, due to axonal destruction by Wallerian degeneration, and typically last >6 months.
  - **Ethyl alcohol:** Concentration required for neurolytic effects is 45% to 100%. 50% concentration is most common for neurolysis to minimize side effects.
- Complications/side effects:
  - Dysesthesias: Pain in the distribution of the sensory component of the blocked nerve. Reported incidence is 10% to 30% and may last from weeks to months. More common when mixed nerves are injected; less frequent with pure motor nerve and motor point blocks.
  - Muscle pain/tenderness
  - Muscle weakness: This may be "permanent," potentially leading to deformity
  - Transient swelling/induration/or nodule formation within the muscle
  - DVT related to decreased motor activity potentially leading to venous stasis
  - Sprains
  - Skin sloughs: May occur superficial to injection site and more common with phenol
    - Phenol can cause serious systemic reactions if injected intravascularly, including convulsions, CNS depression, and cardiovascular collapse. The usual dose of phenol is well below its lethal range: 8.5 g phenol can be a lethal dose; thus, limit dose at single session to 20 to 30 mL of 5%.
  - Alcohol has little to no systemic side effects when injected intravascularly.

Chemodenervation With Botulinum Toxin

GENERAL

- There have been numerous publications and evidence-based reviews demonstrating the safety and efficacy of the botulinum type A toxins for the management of spasticity.
- A family of neurotoxins with seven serotypes (A–G) produced by *Clostridium botulinum* bacteria.
- Botulinum toxin is currently the most widely used treatment for focal spasticity and avoids the generalized weakness and sedation accompanying oral medications.
- In the United States, there are three botulinum type A toxins FDA-approved for clinical treatment of spasticity.
  - OnabotulinumtoxinA (Botox®)
  - IncobotulinumtoxinA (Xeomin®)
  - AbobotulinumtoxinA (Dysport®)
- One type B toxin: RimabotulinumtoxinB (Myobloc®) is FDA-approved for cervical dystonia, not spasticity.
- As of 2024, there are two other botulinum toxin products, prabotulinumtoxinA-xvfs (Jeuveau®) and daxibotulinumtoxinA (Daxxify®), but are not FDA-approved for spasticity.

MECHANISM OF ACTION

- All seven serotypes affect the neuromuscular junction (NMJ) where they produce denervation by blocking the presynaptic release (but not the production) of acetylcholine (ACh) primarily at the terminal portion of a motor nerve.
- Following injection, the toxin is actively taken up by the nerve terminal. All serotypes affect some portion of the SNARE complex, which has three critical components: Synaptobrevin, SNAP-25, and syntaxin. The net effect is to prevent exocytosis of ACh-containing vesicles into the nerve terminal cleft and create an effective chemical denervation.
  - Type A, C, and E cleave SNAP-25
  - Type B, D, F, and G cleave synaptobrevin
  - Type C also cleaves syntaxin
- Toxin is active at the peripheral cholinergic nerve terminals where it inhibits the release of ACh and interferes with the uptake of cytoplasmic ACh.

- Primarily acts at the NMJ but can affect other cholinergic sites including the pre- and postganglionic sites of the autonomic nervous system.

CONTRAINDICATIONS
- Known sensitivity to botulinum toxin
- Presence of infection at the planned injection site

WARNINGS AND PRECAUTIONS
- Distant spread of toxin effect. The FDA has issued a boxed warning on all botulinum toxins regarding the risk of adverse events when the effects of the toxin spread beyond the site where it was injected, including dysphagia and respiratory depression.
- Concurrent treatment is suggested with an aminoglycoside or spectinomycin antibiotics.
- Use with preexisting neuromuscular disease (amyotrophic lateral sclerosis [ALS], myasthenia gravis, Lambert–Eaton myasthenic syndrome, peripheral motor neuropathic disease). These patients are at increased risk for severe reactions, such as dysphagia or respiratory depression, from therapeutic doses.
- Potential corneal exposure or ulceration may occur in patients treated for blepharospasm.
- Safety during pregnancy and lactation has not been established, and botulinum toxins are classified as Category C.

ADMINISTRATION AND DOSING
- One unit of botulinum toxin is equal to the LD_{50} (median lethal dose) for the injection of toxin into an 18-g female Swiss-Webster mouse.
- There is no simple or direct formula for conversion of doses between different toxins.
- Electromyography (EMG), electrical stimulation, and US guidance are all used for muscle localization.
- Dose-dependent response: More toxin load per muscle results in greater clinical weakness.
- General considerations for a spasticity treatment plan:
  - Usual dosage of botulinum toxin depends on muscle size, residual function, size of patient, and degree of spasticity. Consideration is needed regarding the potential harm from weakening of the injected and nearby muscles.
  - Current FDA-approved dosing for adults is 400 units for onabotulinumtoxinA and incobotulinumtoxinA, and 1,000 units for abobotulinumtoxinA. Note: Units of the various toxin products are NOT interchangeable, and the dosing for each toxin is unique.
  - Dosing can be increased with subsequent injections.
  - Experienced clinicians safely use much higher doses with repeat injections.
  - In children, doses of 4 to 8 units/kg are commonly used (400 units considered as maximum body dose).
- As a general rule, reinjections should occur after 3 months.
- Adjunctive treatment should be used in conjunction with botulinum toxins to maximize outcomes, and referral to therapy should be considered.

ONSET AND DURATION OF EFFECTS
- Onset of action usually occurs within 24 to 72 hours, but up to 7 days is not uncommon.
- 🕮 Peak effect: 4 to 6 weeks. Effects typically last 2 to 6 months.
- **Rule of 3's:** "3 days for initial effect, 3 weeks for peak effect, and 3 months duration."

SIDE EFFECTS (USUALLY BENIGN)
- Unwanted weakness in injected or adjacent muscles (localized)
- Hematoma/bruising/local erythema or swelling
- Flu-like syndrome with headache, nausea, fatigue, and general malaise
- Dysphagia may occur from cervical injection (short-lived)
- Nerve trauma
- Pain/soreness

- Antibody formation is not associated with all toxins and may be decreased by maintaining an interval between injections of at least 3 months and keeping doses as low as possible (use smallest effective dose). Antibody formation may be a source of secondary nonresponse.

INTRATHECAL TREATMENTS

Intrathecal Baclofen Pump

GENERAL

- An intrathecal baclofen (ITB) pump allows direct delivery of baclofen into the cerebrospinal fluid (CSF) in the intrathecal space. This makes it possible to achieve a high concentration of baclofen localized to the spinal cord while reducing untoward CNS effects associated with higher oral doses of baclofen (i.e., sedation).
- The ratio of baclofen concentration at the level of the spinal cord for intrathecally administered baclofen compared with orally administered baclofen is approximately 100:1.
- Cervical: Lumbar ratio for drug concentration is 1:3, but the exact effects depend upon the level that the catheter tip is placed. Higher placement may allow for benefit in both the upper and lower extremities. There have been reports of effective catheter placement as high as C4.
- Components of the system include the pump and reservoir, which are implanted subcutaneously in the abdominal wall, and a catheter, which is surgically placed into the intrathecal space.
- An implantable infusion system with a programmable battery-powered pump is used to store and deliver the baclofen via an electronically entered schedule or timetable.
- ITB pumps are refilled on an intermittent basis via transcutaneous injection performed with the assistance of a superficial injection template and a computerized programmer. The frequency of refill depends upon infusion dosing rate and size of the pump reservoir (20 or 40 mL). Currently available pumps have a battery life of 80 months.
- Dose and function parameters are adjustable transdermally using a computerized programmer.
- Dose schedule can be set to schedule a continuous infusion or to vary the dose through the day depending upon clinical need.
- The pump will emit an audible tone prior to the reservoir running out of medication.

INDICATIONS

- Patients with generalized, diffuse spasticity who are intolerant to or lack response to more conservative treatments (oral agents, nerve blocks, etc.).
- Care must be taken to evaluate the patient in regard to the presence of useful spasticity. If the patient has functional use of their spasticity, then an ITB pump may not be appropriate.
- Used for patients with spinal forms of spasticity, SCI or MS, and acquired TBI, stroke, and CP, provided there is some preserved function below the level of the lesion.
- Prior to implantation of an ITB pump, patients are generally given a trial of ITB through a single bolus dose or via continuous infusion through a lumbar puncture/percutaneous catheter. Multiple trials with increasing doses may be tried, usually with an initial starting dose of 50 μg followed by trials at 75 and 100 μg as needed to demonstrate clinical benefit.
- If there is significant reduction of tone, frequency, or severity of spasms with the trial dose, without an adverse reaction, the patient is a good candidate for the pump. Response is usually measured by a reduction in the Ashworth score of preselected muscles.
- Initial pump infusion dosing can be estimated based upon the trial dose required to achieve a clinical response. If the ITB trial is done by bolus, a common method is to double the clinically effective trial dose and use this amount as the initial 24-hour infusion dose. Continuous ITB trial has the advantage of allowing the patient to experience the effects of ITB at various doses and potential adverse effects by reaching steady states of ITB mimicking permanent pump placement.
- ITB pumps require a very compliant patient and/or caregivers given the need for regular monitoring of the pump and refilling of the reservoir (usually every 3–5 months). Noncompliance places the patient at risk for significant complications, including baclofen withdrawal, which may be fatal.

SIGNS/SYMPTOMS OF BACLOFEN OVERDOSE

- Hypotonia/severe weakness
- Drowsiness/somnolence
- Lightheadedness
- Nausea/vomiting
- Bradycardia
- Hypotension
- Seizures
- Respiratory depression
- Loss of consciousness progressing to coma
- These are usually dose-related and often resolve with dose reduction.
- The anticholinesterase physostigmine (2 mg IV) may be given to reverse the respiratory depression caused by baclofen overdose.

🕮 SIGNS/SYMPTOMS OF BACLOFEN WITHDRAWAL

- Pruritus without rash
- Fever
- Nausea
- Dizziness
- Hyperthermia
- Insomnia
- Altered mental status
- Hallucinations
- Seizures
- Exaggerated rebound spasticity and muscle rigidity that can lead to rhabdomyolysis and multiple organ system failure

PROBLEMS ASSOCIATED WITH INTRATHECAL BACLOFEN PUMP MECHANISMS

- Catheter dysfunction (dislodging, migration of the tip, kinking, disconnection from the pump apparatus, blockage)
- Pump failure
- Infection
- Errors in baclofen dosage/programming errors
- Battery failure
- Skin breakdown over pump implantation site
- Spinal headache due to CSF leakage around the catheter

🕮 MANAGEMENT OF INTRATHECAL BACLOFEN WITHDRAWAL/OVERDOSE

INTRATHECAL BACLOFEN WITHDRAWAL/LOSS OF EFFICACY

Patients with previously controlled spasticity presenting with worsening tone should undergo prompt evaluation to determine if the etiology is related to pump malfunction. If not related to pump malfunction, increased spasticity may be related to infection, pressure injury, constipation, gallstones, ileus, or other pain generators. Evaluation of pump function may include:

- Pump interrogation/review of logs
- Program bolus dose to look for anticipated improvement in tone
- Confirm actual pump reservoir against expected
- Perform diagnostic catheter access port aspiration to check for catheter integrity
- Pump/catheter imaging: May include lateral and anteroposterior (AP) x-rays, fluoroscopic studies, or CT myelogram

If the patient has confirmed pump malfunction and is experiencing withdrawal, treatment includes:

- Hospital admission for monitoring of cardiopulmonary and renal status
- Neurosurgical consultation for correction of pump malfunction

- Bolus administration of ITB via lumbar puncture for severe withdrawal
- Medication interventions including baclofen, IV benzodiazepines, and cyproheptadine
- Intravenous (IV) fluids

INTRATHECAL BACLOFEN OVERDOSE

Patients with ITB overdose may present with nausea and vomiting, dizziness, respiratory depression, hypothermia, somnolence, and/or seizures. Similar to ITB withdrawal, a comprehensive history and physical examination should be performed to rule out other causes of presentation including sepsis, hypoglycemia, electrolyte imbalance, seizure disorder, and intracranial hemorrhage. Management may include:

- Hospital admission for monitoring of cardiopulmonary status
- Reduction or temporary interruption of ITB delivery by pump reprogramming
- Consideration of CSF drainage via catheter aspiration port (CAP) aspiration
- Intubation for airway protection in severe cases
- Monitoring for rebound withdrawal

Surgical Treatments

ORTHOPEDIC PROCEDURES

- Allow for potential improvement in passive ROM (PROM) in joints affected by spasticity.
- Usually irreversible and require significant preprocedure planning
- Realistic goal setting is vital and must include all parties: Patient, family, and healthcare providers.
- Preprocedure dynamic EMG, botulinum toxin, and/or nerve blocks may be beneficial in both planning specifics of the surgical intervention and in developing realistic goals.
- In addition to specific muscle spasticity, the clinical exam should attempt to delineate any muscle co-contraction and out-of-phase muscle firing.
- ***Tendon lengthening procedures:***
  - Tendon release or lengthening
  - Step-cut (Z-plasty) lengthening procedures, commonly performed on the Achilles tendon
  - Tenotomy: Commonly performed on the hamstrings and iliopsoas
  - Myotomy: Adductor myotomies commonly performed in the setting of recurrent spasticity associated with hip dislocation
- ***Tendon transfer procedures:***
  - 🕮 Split Anterior Tibial Tendon Transfer **(SPLATT):** A commonly used tendon transfer procedure used to treat equinovarus deformity of the foot. Spasticity in the tibialis anterior (TA) muscle provides an inverting and supinating force on the foot, while spastic gastrocnemius and soleus muscles provide a plantar flexing force.
    - During the SPLATT procedure, the TA muscle is split distally along its length, and the distal end of the lateral half of the muscle is attached to the cuboid and third cuneiform bones. This transfer creates a balancing eversion force without significant loss of dorsiflexion force and is usually done in conjunction with an Achilles lengthening to address the equinus component of the deformity.
    - Postoperatively, the joint is generally stabilized with a short leg cast or rigid boot for 2 to 6 weeks, followed by a rigid ankle–foot orthoses (AFO) for an additional 6 weeks. This allows for 12 weeks of protection to allow the transferred tendon and its surgical insertion to heal and protect from inadvertent stretch.
  - Multiple other tendon transfers are commonly performed: Brachioradialis transfer to the extensor side of the forearm; flexor hallucis longus and digitorum longus transfer to the os-calcis.

NEUROSURGICAL PROCEDURES

- In general, neurosurgical procedures involve a surgical sectioning at the level of the peripheral nerve, nerve rootlets, spinal tracts, or spinal cord.

- Intrathecal drug administration is performed including ITB pump as discussed previously.
- Central electrical stimulators used, including epidural spinal cord stimulation and cerebellar stimulation.
- Rhizotomy: Surgical intervention at the level of the nerve rootlets. This may be a general surgical sectioning of both motor and sensory rootlets, a sectioning of only sensory rootlets (selective dorsal root rhizotomy), or radioablative intervention of rootlets (radiofrequency rhizotomy).
  - Selective dorsal root rhizotomy attempts to ablate the most pathologic sensory rootlets as noted with intraoperative electrophysiologic studies. The goal of this procedure is to modulate afferent sensory input and reset muscle spindles to decrease spasticity with the minimum loss of sensation and preservation of motor output. More commonly used as a potential therapy for children with CP; use is not as well-defined in the adult population.
  - Numbness may result from this procedure.
- Peripheral neurectomy
- Neuroablative procedures
- Spinal cord surgical procedures are much more aggressive procedures and reserved for severely spastic patients who have failed all other attempted interventions.
  - Cordotomy: Sectioning portions of the spinal cord; this may be effective, but high risk exists for loss of bowel and bladder function
  - Myelotomy: Severing tracts of the spinal cord

MOVEMENT DISORDERS

- Movement disorders refer to a group of CNS neurodegenerative diseases associated with involuntary movements or abnormalities of skeletal muscle tone and posture.
- The predominant area of involvement in movement disorders is the extrapyramidal system, especially the basal ganglia, but other pathways and structures can be affected. The basal ganglia are primarily inhibitory and influence direction, amplitude, and course of movement.
- Cerebellar function also plays a role in movement disorders and affects rapid corrections of gross motor movements and coordination of movements.
- Typically, primary movement disorders are not associated with weakness or sensory loss.

🕮 GENERAL DEFINITIONS

Chorea

From the Greek word "to dance," chorea refers to brief, rapid, forceful, and dysrhythmic flinging of the limbs (or other body parts). Choreiform movements are nonstereotyped, unpredictable, jerky movements that interfere with purposeful motion. The most familiar, generalized form is found in Huntington's disease (HD).

Athetosis

Slow, writhing, involuntary movements, and inability to maintain the position of a limb or any other body part. Slower than choreiform movements and less sustained than dystonia, it usually involves the face and distal upper extremities. Etiology may be idiopathic but can be related to another neurologic disease, such as stroke, tumor, or Wilson's disease.

Hemiballismus

Rare disorder characterized by sudden onset of violent, involuntary movements on one side of the body (unilateral and contralateral to the lesion), mainly of the arm, but can also involve the leg. It is usually associated with hemorrhage or infarction of the contralateral subthalamic nucleus (which regulates the globus pallidus).

Tics

Sustained, nonrhythmic muscle contractions that are rapid and stereotyped, often occurring in the same extremity or body part during times of stress.

Akathisia

Akathisia (Greek meaning "not to sit") refers to an extrapyramidal movement disorder caused by reversible motor restlessness accompanied by a sensation of unpleasant inner tension or anxiety, often confused with psychotic agitation. The most common cause is iatrogenic, as a complication of antidopaminergic drugs such as antipsychotics. Incidence varies widely but occurrence is higher with first-generation antipsychotics than with second-generation antipsychotics (Bratti et al., 2007).

Ataxia

Defined as a lack of muscle coordination that is not due to paresis, alteration in tone, loss of postural sense, or the presence of involuntary movements. Ataxia is the most important sign of cerebellar disease, but it can also indicate problems in the dorsal columns or peripheral nerves (sensory ataxia) or vestibular system (vestibular ataxia).

- **Sensory ataxia:** Due to loss of proprioception
- **Vestibular ataxia:** Associated symptoms include vertigo, nausea, and vomiting.
- **Cerebellar ataxia:** Associated neurologic deficits include dysmetria, asynergia, and dysdiadochokinesia.
  - *Dysmetria* is a disturbance in the trajectory or placement of a body part during active movement, resulting in either hypometria (undershooting of the target) or hypermetria (overshooting of the target). It is commonly the result of lesions to the cerebellum (caused by stroke, MS, ALS, tumors).
  - *Asynergia* or dyssynergia refers to difficulties within the sequence and speed of the components of movement.
  - *Dysdiadochokinesia* is difficulty with repetitive or fine movement.

Tremors

Rhythmic, oscillatory movements of a body part. They are classified according to the circumstances during which the tremor occurs (Bhatia et al., 2018).

- ***Resting tremor:*** Occurs at rest, as seen in Parkinson's disease (PD). Involuntary movement usually affects the hands and fingers.
- ***Postural tremor:*** Occurs during maintenance of a specific body posture.
- ***Action tremor:*** Occurs during voluntary movement from one point to another. Often a sign of cerebellar dysfunction (further described below). An intention tremor is the exacerbation of an action tremor at the end of the action and is a component of the action tremor.
- ***Essential tremor:*** Occurs when maintaining the position of a body part against gravity (further described below).

ESSENTIAL TREMOR

- Most common tremor, usually affects the hands and the head.
- Posture-maintaining tremor associated with sustained muscle contraction or stress.
- Incidence increases with age, affecting 0.04% of people under age 20 and 3% of people over age 80 (Song et al., 2021).
- Benign (usually tremor is the only problem and does not lead to other neurologic impairment), although in 10% of patients can evolve into Parkinson's.
- May be familial (often familial, however no specific gene identified) or sporadic.
- Tremor may attenuate or disappear with intake of small amounts of alcohol.
- Treatment: Beta-blockers (e.g., propranolol), primidone, anticholinergic drugs, benzodiazepines, MRI-guided focused US thalamotomy, deep brain stimulation (DBS) to the

thalamus, and wearable devices such as the Cala Trio®, which uses transcutaneous afferent patterned stimulation (Haubenberger & Hallett, 2018; Okelberry et al., 2024).

ACTION TREMOR

- Occurs with movement from point to point, often at the end of a purposeful movement; it is evaluated with the "finger-to-nose" test.
- Amplitude of tremor increases as extremity approaches target.
- Cerebellar tremor is an example of an intention tremor and is caused by damage to the cerebellum (seen in stroke, MS, neurodegenerative diseases, or alcoholism).
- This type of tremor is minimally treatable with medications or surgery, but wrist weights can be helpful.

Myoclonus

- Characterized by sudden, jerky, irregular, or periodic involuntary contractions of a muscle or group of muscles.
- Usually caused by a disturbance to the CNS, possibly due to an overexcitability of the parts of the brain that control movement. Often seen after anoxic episode (Lance-Adams syndrome). Other etiologies include hyponatremia, renal/hepatic failure, hypoglycemia, or toxins.
- There are many classification systems for myoclonus:
  - Reflex myoclonus: Stimulus-sensitive myoclonus, occurs with volitional movements, muscle stretch, or superficial stimuli-like touch
  - Nonspontaneous myoclonus: Nonstimulus-sensitive, occurs at rest
  - Positive myoclonus: Involuntary movement with sudden muscle contracture
  - Negative myoclonus: Involuntary movement with relaxation of the muscle
- Treatment depends on the subtype of myoclonus and includes oral medications (levetiracetam, clonazepam, and valproic acid) or chemodenervation with botulinum toxin.

Dystonia

Sustained muscle contractions producing abnormal repetitive, twisting movements of variable speed that lead to abnormal movements or posturing.

- While once considered primarily idiopathic, numerous genetic mutations have been linked to the condition.
- Can also result from trauma or a pharmacologic agent, especially antipsychotic drugs (Stephen, 2022)
- May be generalized (involving trunk and two or more other body parts), focal (one specific body part), or segmental (two or more contiguous body parts; Stephen, 2022)
- "Primary" dystonias usually first manifest as partial dystonia in childhood or adolescence and progress to generalized. They are caused by genetic mutations (mostly autosomal dominant, while some are recessive; Stephen, 2022; Tanabe et al., 2009).
- Oral medications are generally not very effective in controlling dystonia, but small doses of levodopa may be effective for dopa-responsive dystonia, which represents a very small subset of the population with dystonia.
- Physical therapy (PT) can be beneficial for focal dystonia, especially in combination with other treatments.
- Prognosis in primary dystonia is dependent on age of onset. Younger age ⟶ more likely to progress to generalized disabling dystonia.
- Course can be very variable with spontaneous remissions and exacerbations.
- Intramuscular injection of botulinum toxin is the most effective treatment of focal dystonia.
- DBS can be very effective in patients with primary dystonias, especially those associated with DYT-1 genetic mutations (Stephen, 2022).
- Cervical dystonia is the most common type of focal dystonia.

  - Subtypes include torticollis, laterocollis, anterocollis, and retrocollis (see the following table for the muscles involved in each subtype).
- Blepharospasm is the second most common focal dystonia. One eye may be affected initially, but both eyes ultimately become involved.

SUBTYPES OF CERVICAL DYSTONIA	PRIMARY MUSCLES AFFECTED
Torticollis (head turning; please refer to Chapter 10, Pediatric Rehabilitation, for elaboration of torticollis)	Ipsilateral splenius capitis Contralateral sternocleidomastoid Splenius cervicis Levator scapulae Inferior oblique longus capitis
Laterocollis (head tilt)	Splenius capitis Ipsilateral scalene complex Levator scapulae Longissimus
Retrocollis (neck extension)	Bilateral splenius capitis Posterior capitis and longissimus
Anterocollis (neck flexion)	Bilateral sternocleidomastoid

RESTLESS LEGS SYNDROME

General

- Dysesthetic sensations in the lower > upper extremities
- Patients report a strong, sometimes irresistible urge to move their legs. Usually not painful but can be bothersome.
- Often associated with sleep and is worse with inactivity. The urge to move or unpleasant sensations begin or worsen during periods of rest or inactivity such as lying or sitting.
- These sensations are partially or completely relieved by movement such as walking or stretching.
- The sensations are worse in the evening than during the day.
- Affects 10% to 15% of the population. Pathogenesis is unclear.
- Possibly due to deficiency of dopamine D2 receptors and reduced availability of serotonin transporter in the brain. Other potential mechanisms include sympathetic hyperactivity or underactivity of GABA systems.
- Types:
  - Primary restless legs syndrome (RLS): Idiopathic CNS disorder
  - Secondary RLS: Most commonly due to iron deficiency, end-stage renal disease, or peripheral neuropathy

TREATMENT

- Dopaminergic agents: Bedtime low dose of carbidopa-levodopa (Sinemet®), ropinirole (Requip®), rotigotine (Neupro®), and pramipexole (Mirapex®). Most patients with RLS show at least an initial response to dopaminergic agents (Lv et al., 2021).
- Gabapentin enacarbil extended-release (Horizant™; Kim & Deeks, 2016)
- Benzodiazepines: Clonazepam (Klonopin®; Lv et al., 2021)

🕮 PARKINSON'S DISEASE (TABLE 12–3)

- PD is the second most common neurodegenerative disease after Alzheimer's disease (Kouli et al., 2018). Symptoms do not present in most individuals with PD until up to 70% of the dopamine-producing cells (in the substantia nigra) are gone (Dauer & Przedborski, 2003; Surmeier, 2018).

TABLE 12–3 Parkinson's Disease

General	• Idiopathic PD is a disorder of the basal ganglia due to loss of cells in the SN and LC, where dopamine is produced, and degeneration of the nigrostriatal pathway (from SN to the corpus striatum). This results in a ↓ in dopamine content in the corpus striatum. • Microscopically, intracytoplasmic eosinophilic inclusions called Lewy bodies are found in damaged cells. • Dopamine depletion affects the direct and indirect pathways in the striatum that modulate voluntary movements through thalamo-cortical output.
Epidemiology	• Greatest risk factor for PD is advanced age, with increased incidence after age 65 (Willis et al., 2022) • Male to female ratio of 1.4:1.0 (Armstrong & Okun, 2020) • Prevalence: 572/100,000 in people age >45; over 1 million individuals in the United States • Incidence: – 50–70/100,000 per year in people age >45 – 108–212/100,000 per year in people age >65 • Disease is progressive, with progressive motor disability over time and development of nonmotor complications in late stages, including dementia, psychosis, and autonomic dysfunction. • Life expectancy, however, can be near-normal, despite the motor and nonmotor disability. • EOPD or YOPD—diagnosis before age 50. Occurs in 10%–20% of people diagnosed. It is a subtype characterized by slower progression of disease, increased rate of dystonia and levodopa-induced dyskinesia, and less motor and cognitive disability.
🕮 Signs/ Symptoms	**PD symptoms can be differentiated into motor and nonmotor symptoms.** **Motor Symptoms** • **Resting tremor** ("pill-rolling") usually at 3–5, 4–6 Hz: – Most common symptom, affecting two-thirds of patients, usually affecting hands and feet unilaterally – Can have postural tremor • **Bradykinesia/hypokinesia** (slowness of movements) • **"Cogwheel" rigidity** (tremor superimposed on muscular rigidity) • **Postural instability/loss of postural reflexes** (with tendency to fall to the side or backward) • Micrographia, difficulties cutting food, feeding, dressing • Masked facies (hypomimia—expressionless face) • Gait in a festinating (shuffling) gait pattern with decreased arm swing, short steps, marked flexion of the trunk during ambulation and standing (known as camptocormia), narrow base of support. • PISA syndrome—reversible lateral bending of the trunk with tendency to lean to one side • Postural instability/loss of postural reflexes (with tendency to fall to the side or backward) • FOG phenomena—transient inability to perform or restart certain task or transient inability to move **Nonmotor Symptoms** • Dementia (seen in one-third of patients) • Behavioral disorders: Depression, apathy, anhedonia. Depression is seen in one-third of patients • Sensory: Anosmia, impaired visual acuity, impaired visual contrast perception, impaired depth perception • Autonomic dysfunction: Orthostatic hypotension, constipation, urinary and sexual dysfunction • Sleep disorders: REM sleep behavior disorder, vivid dreams, restless legs syndrome
Medical Treatment Goals	**Goals:** ↑ dopamine action 1. **Carbidopa/levodopa combination:** Mainstay of PD treatment. Levodopa is the metabolic precursor to dopamine given with carbidopa, which blocks metabolism of L-dopa to dopamine. – Sinemet®—carbidopa + levodopa immediate-release – Rytary®—carbidopa/levodopa extended-release

(*continued*)

TABLE 12–3 Parkinson's Disease (*continued*)

Medical Treatment Goals (*cont.*)	– Stalevo®—carbidopa/levodopa + entacapone (COMT inhibitor) – Duopa®—parenteral infusion – Side effects of levodopa include motor fluctuations with dyskinesias, orthostatic hypotension, nausea, confusion, and hallucinations. ■ Treatment with levodopa over time usually results in response fluctuations (characterized by "on/off" periods) that vary in levels of unpredictability and can significantly impact quality of life. Exact mechanism is unknown. ■ Treatment of "off" periods include adjustments of dose, frequency, and timing, as well as adjunct therapies to extend duration of levodopa effect with COMT inhibitors and MAO inhibitors (Rota et al., 2022) 2. **Dopamine receptor agonists:** – Ropinirole (Requip®) – Pramipexole (Mirapex®) – Rotigotine (Neupro® patch) – Behavioral side effects can also occur with dopamine agonists, including impulse control disorders that can include compulsive gambling and sexual behaviors. 3. **Amantadine (Symmetrel®):** Antiviral agent; acts primarily by potentiating the release of endogenous dopamine. Also has mild anticholinergic activity. – Somewhat inhibits dopamine reuptake – Glutamate receptors blocking – Long-acting amantadine (Gocovri®) is effective in levodopa-induced dyskinesias. 4. **Anticholinergic agents (muscarinic receptor antagonists):** – Occasionally useful in relieving tremor; however, rarely used secondary to side effects, including confusion, sedation, worsening constipation, and urinary dysfunction – Trihexyphenidyl (Artane®) – Benztropine (Cogentin®) 5. **Inhibitors of dopamine metabolism:** – Selective monoamine oxidase B (MAO-B) inhibitors: ■ Has mild symptomatic effect in early PD ■ May reduce the oxidative damage in the SN and slow the disease progression ■ No evidence to support the earlier notion that these agents are neuroprotective and delay the natural progression of the disease ■ Selegiline (Eldepryl®) ■ Rasagiline (Azilect®) ■ Safinamide (Xadago®) **Note:** The MAO-B isoenzyme is the predominant form (compared with MAO-A) in the striatum and is responsible for the majority of the oxidative metabolism of dopamine in the striatum. – COMT inhibitors: ■ Inhibit metabolism of dopamine and its precursor levodopa in the liver, GI tract, and other peripheral organs ■ Used as adjunct treatments to levodopa-carbidopa; may allow a reduction on the levodopa dosage. Must be taken with levodopa to have an effect. ■ Tolcapone (Tasmar®) ■ Entacapone (Comtan®) ■ Opicapone (Ongentys®) ■ Carbidopa/levodopa + entacapone combination tablets (Stalevo®)
🕮 Advanced Therapies	• Advanced therapies are used to treat Parkinson's symptoms (rigidity, dyskinesias, tremor) in patients with medication-responsive motor symptoms but have complications such as off periods or dyskinesias that are refractory to medication adjustments (Armstrong & Okun, 2020). • Patients with dementia or significant psychiatric/behavioral symptoms are not candidates for these treatments. • 🕮 DBS (Armstrong & Okun, 2020): Considered the preferred surgical treatment for disabling PD-related tremor and levodopa-related motor complications. – 🕮 Surgical placement of unilateral or bilateral electrodes into the subthalamic nucleus or globus pallidus interna. The electrode is connected to a battery placed in the chest and a pulse generator which delivers high-frequency stimulation of varying dosage and frequency. It is purported to work by resetting abnormal firing patterns in the brain.

(*continued*)

TABLE 12–3 Parkinson's Disease (*continued*)

🕮 Advanced Therapies (*cont.*)	– The procedure is associated with fewer complications than thalamotomy and has replaced it as the surgical procedure of choice. – *Subthalamic nucleus DBS* effectively reduces tremor, rigidity, and bradykinesia. It allows for a reduction in antiparkinsonian medications by at least half. Currently, it is the most common surgical procedure for PD. – *Pallidal DBS* controls all the cardinal symptoms of PD. Implantation of a lead is in the globus pallidus interna. – *HFU* is FDA-approved to treat specific symptoms of PD; however, it is not widely used (with DBS remaining the standard procedure used for medication-refractory PD symptoms). It can be used for sonification of the thalamus, globus pallidus, subthalamic nucleus, or pallidothalamic tract. **Other Treatments:** • PT and rehabilitation medicine are effective in improving motor function and gait and can also delay disease progression. • Regular exercise can significantly delay disease progression especially in early stages of the disease. • Chemoneurolysis with botulinum toxin to treat muscle overactivity to address tremor and rigidity, but is rarely used in clinical practice because of possible weakness that can limit motor dexterity. • rTMS—noninvasive procedure that delivers repeated magnetic pulses to a specific brain area through a stimulation coil placed over the scalp. Evidence suggests rTMS improves motor symptoms for patients with PD. • tDCS—another mode of noninvasive brain stimulation, whereby a direct current is applied via surface electrodes. • Focused ultrasound thalamotomy uses focused ultrasound beams under MRI guidance to burn the thalamus. This effectively improves tremors but can only be used unilaterally due to side effects such as worsening speech and balance.
Rehab Treatment	• Exercise has been shown to offer protective benefits from onset of PD and significantly affect the natural progression of PD. • Training in rhythmic pattern to music as well as verbal, tactile, and visual cueing may help overcome the effects of freezing and festination. • Education to caregiver and support for community assistance programs • 🕮 Swallowing evaluation to evaluate for oropharyngeal dysphagia • Speech therapy for dysarthria and dysphonia • Hypophonia can be overcome by training the patient in voice enhancement. Treatments include LSVT and PLVT. • Assessment of degree of rigidity, bradykinesia, and decrease in manual dexterity and how they affect ADLs • Adaptive equipment can be provided when deficits in upper extremity control limit efficient and safe function. Examples of adaptive equipment to improve feeding: – Plate-guards or specialized dishes – Weighted or large-handled cups and utensils – Swivel fork and spoons – Velcro® or zipper closures instead of buttons may improve dressing. • Gait evaluation should be done, including speed and distance. • Assessment of fine motor tasks, such as writing • Evaluation of cognitive function if clinically indicated • Vibrating utensils for tremor reduction • Social isolation is common. • Walking becomes impaired as the disease progresses. • Wheeled walkers are preferred to standard walkers with visual light beams and/or sound features serving as cues (e.g., the U-Step Walker®). • Fall prevention; consider home assessment to modify environmental factors (e.g., loose rugs).

ADL, activities of daily living; COMT, catechol-*O*-methyltransferase; DBS, deep brain stimulation; EOPD, early-onset Parkinson's disease; FDA, Food and Drug Administration; FOG, freezing of gait; GI, gastrointestinal; HFU, high-intensity focused ultrasound; LC, locus coeruleus; LSVT, Lee Silverman Voice Treatment; MAO-B, monoamine oxidase B; PD, Parkinson's disease; PLVT, pitch limiting voice treatment; PT, physical therapy; REM, rapid eye movement; rTMS, repetitive transcranial magnetic stimulation; SN, substantia nigra; tDCS, transcranial direct current stimulation; YOPD, young onset Parkinson's disease.

Differential Diagnosis of Parkinson's Disease

- Drug-induced parkinsonism: Exposure to drugs, such as neuroleptic agents (first generation, e.g., haloperidol; second generation: Olanzapine, iloperidone, lurasidone, etc.), metoclopramide (Reglan®), reserpine, amiodarone (rare), and lithium (Feldman et al., 2022; Orr & Ahlskog, 2009; Wisidagama et al., 2021)
- Toxin-induced parkinsonism (1-methyl-4-phenyl-1,2,3,6-tetrahydropyridine, manganese, carbon monoxide)
- Cerebrovascular accidents/multiple lacunar strokes
- Brain tumors
- Dementia pugilistica (posttraumatic parkinsonism): Parkinsonism associated with repeated trauma to the head.

Parkinson's Plus Syndromes

- Signs of parkinsonism plus additional signs of neurodegenerative lesions. May mimic PD in the first 1 to 2 years after clinical diagnosis but are poorly responsive to levodopa therapy and have an overall worse prognosis than PD with-more rapid disease progression.
- Parkinson's plus syndromes include progressive supranuclear palsy (PSP), multiple system atrophy (MSA), CBD, and diffuse Lewy body disease.

Progressive Supranuclear Palsy

- Supranuclear ophthalmoparesis is one of the hallmark signs, manifested with a paralysis of vertical gaze.
- The earliest and most disabling symptom of PSP usually relates to gait and balance impairment, resulting in patients frequently falling and sustaining injuries. In contrast to short, shuffling steps in PD gait, patients with PSP have a stiff broad-based gait with knees and trunk extended, arms abducted, and a tendency to fall backwards as a result.
- Marked change in personalities. Affected individuals appear apathetic and may appear depressed; they may have monotonous speech. Dysarthria and dysphagia may also occur early in the course of the disease, as well as cognitive deficits.

Multiple System Atrophy

- Clinically characterized by a combination of parkinsonism, cerebellar dysfunction, and autonomic dysfunction (orthostatic hypotension, urinary dysfunction, constipation, salivary and thermoregulatory dysfunction)
- Pathophysiology involves damage to the oligodendrocytes by accumulation of the protein alpha-synuclein.
- Motor types, depending on the prominent clinical presentation, can be divided into MSA-P (parkinsonian type) and MSA-C (cerebellar type). MSA-C is characterized by ataxia and other cerebellar findings including a wide-based gait and cerebellar oculomotor disturbances. MSA-P is characterized by parkinsonian features with autonomic dysfunction.

Huntington's Disease (TABLE 12–4)

- In a small percentage of patients with HD, hypokinesia and rigidity are the presenting features (similar presentation to PD). This is known as the *Westphal variant* and tends to have an earlier age of onset.

TABLE 12–4 Huntington's Disease (HD)

Description	• Hereditary disease (autosomal dominant) • Abnormal movements (chorea) due to heightened sensitivity of striatal dopamine receptors • The disease predominantly strikes the striatum. Atrophy of the caudate nucleus can be seen on neuroimaging studies (MRI). • Neurochemically, there is marked ↓ of GABA throughout the basal ganglia; there is also ↓ levels of substance P and enkephalins. • Genetics: HD gene (a trinucleotide CAG repeat) identified near the tip of the short arm of chromosome 4 (4p16.3) • Huntington is the abnormal protein product of this gene that accumulates in brain cells.
Epidemiology	• Prevalence in North America and Europe: 4–8 per 100,000 in the general population, significantly higher than that of Asia and Africa (Medina et al., 2022) • Usually begins at age 30–50 years • 10% have juvenile onset (age <20 years) • Death occurs on average 15–20 years after the onset, usually from aspiration pneumonia.
Signs/ Symptoms	**Classic Triad** 1. Chorea/choreoathetosis (hyperkinetic, involuntary, jerky movements) 2. Dementia and personality disorders 3. Family history (dominant inheritance): – Dysarthria – Teeth grinding – Facial grimacing • Difficulty swallowing • Depression • Rigidity can also be a feature but is more common in juvenile-onset illness • Death occurs (on average) 15–20 years after onset (usually from aspiration pneumonia)
Treatment	**Goal: ↓ Dopamine Action** **Vesicular Monoamine Transport 2 Inhibitors (VMAT2 Inhibitors)** • Deplete neurotransmitters' stores of dopamine, serotonin, and other monoamines • Deutetrabenazine (Austedo®)—indicated for treatment of tardive dyskinesia and chorea associated with HD • Valbenazine (Ingrezza®)—approved for the treatment of HD chorea • Tetrabenazine is rarely used because of potential serious side effects, including depression and parkinsonism. **Neuroleptics/Dopamine Receptor Blockers** 1. Haloperidol (effective in suppressing chorea but has extrapyramidal and anticholinergic side effects) 2. Phenothiazines: – Fluphenazine – Perphenazine – Side effect: Tardive dyskinesia **Presynaptic Dopamine Depleters** • Reserpine: Effective in treatment of chorea (side effect—hypotension) • Other agents for treatment of depression, psychosis, irritability: – Clozapine, quetiapine – Fluoxetine and other SSRIs – TCAs – Carbamazepine

GABA, gamma-aminobutyric acid; SSRI, selective serotonin reuptake inhibitor; TCA, tricyclic antidepressant.

CLASSIFICATION OF ATAXIAS (TABLE 12–5)

In 1983, Harding introduced the now widely accepted classification system of ataxias. He divided them into two groups, hereditary and nonhereditary. The hereditary ataxias were further divided into autosomal dominant, autosomal recessive, and X-linked.

TABLE 12–5 Classification of Ataxias

HEREDITARY ATAXIAS
Autosomal recessive ataxias: • Friedreich's ataxia (FA) • Early-onset cerebellar ataxia (EOCA) • Ataxia-telangiectasia (AT) • Ataxia with isolated vitamin E deficiency (AVED) *Autosomal dominant ataxias:* • Spinocerebellar ataxias (SCA) • Dentatorubral-pallidoluysian atrophy • Episodic ataxias (EA) *X-linked ataxias:* • Fragile X-associated tremor ataxia syndrome (FXATS)
NONHEREDITARY ATAXIAS
• Multiple system atrophy, cerebellar type (MSA-C) • Sporadic adult-onset ataxia of unknown etiology • Symptomatic ataxias: – Alcoholism (alcoholic cerebellar degeneration) – Toxins (antiepileptics, lithium, solvents) – Malignancy (paraneoplastic cerebellar degeneration) – Malabsorption (acquired vitamin E, vitamin B_{12} deficiencies) – Immune-mediated ataxias (gluten ataxia, ataxia associated with anti-glutamic acid decarboxylase [GAD] antibodies) – Hypothyroidism – Physical causes (heat stroke, hyperthermia)

Source: Adapted from Goetz CG, Pappert EJ. *Textbook of Clinical Neurology*. W.B. Saunders; 2007, with permission.

Hereditary Autosomal Recessive Ataxias

FRIEDREICH'S ATAXIA

- Caused by a problem with low production of messenger RNA from gene X25
- Prevalence 0.4 to 4.7 per 100,000
- Presents most commonly before age 25 and most likely between ages 10 and 15 years old
- Patients present with progressive ataxia, areflexia of the lower limbs, impaired vibration, or position sense.
- Two-thirds of patients have extensor plantar reflexes, progressive weakness of extremities, oculomotor abnormalities, rare nystagmus, dysarthria, optic atrophy, reduced visual acuity, and sensorineural hearing loss.
- Nonneurologic signs: Scoliosis, pes cavus, and hypertrophic obstructive cardiomyopathy

ATAXIA-TELANGIECTASIA

- Progressive ataxia starting in early childhood, oculocutaneous telangiectasia, and a high incidence of neoplasia

EARLY-ONSET CEREBELLAR ATAXIA WITH RETAINED TENDON REFLEXES

- Differentiated from Friedreich's ataxia by the presence of deep tendon reflexes. May be several different disorders with possible X linkage, as well as some noncongenital sources.
- Prevalence 0.5% to 2.3% per 100,000
- Starts by age 20 years
- Presents with progressive ataxia of gait, stance, and limb movements, dysarthria, and oculomotor disorders, with half having impaired vibration or position sense

Hereditary Autosomal Dominant Ataxias

SPINOCEREBELLAR ATAXIAS

- More than 25 different gene loci have been identified.
- Mutation has been identified as a translated cytosine adenine guanine (CAG) repeat expansion in six types: spinocerebellar ataxia (SCA) 1, 2, 3, 6, 7, and 17.

DENTATORUBRAL-PALLIDOLUYSIAN ATROPHY (DRPLA)

- Found primarily in Japan with a prevalence of 0.2 to 0.7 per 100,000. Mutation identified as a CAG repeat expansion in the DRPLA gene on chromosome 12p13 (Le Ber et al., 2003).
- Average onset 30 years within range from infancy to late adulthood. Ataxia, dysarthria, and progressive dementia are symptoms. When onset is before age 21, myoclonic epilepsy is also seen. Can have psychiatric disturbance, choreic, or dystonic movements.

EPISODIC ATAXIA TYPE I

- Autosomal dominant disorder caused by potassium channel gene on chromosome 12p
- Unknown prevalence presents in early childhood with brief attacks of ataxia and dysarthria associated with interictal myokymia

EPISODIC ATAXIA TYPE II

- Autosomal dominant disorder linked to chromosome 19p. Normally presents during childhood as early as 6 weeks to 30 years of age.
- Different from Type I because of long duration of attacks, lasting hours to greater than a day

X-Linked Ataxias

FRAGILE X-ASSOCIATED TREMOR/ATAXIA SYNDROME

- Fragile X syndrome is the most common inherited form of intellectual disability.
- X-linked dominant disorder commonly due to silencing of the FMR1 gene secondary to expansion of a CGG repeat (Salcedo-Arellano et al., 2020).
- Fragile X-associated tremor ataxia syndrome (FXATS) is an adult-onset fragile X syndrome variant; progressive action tremor and cerebellar ataxia also develop as part of this syndrome.

Nonhereditary Ataxias

MULTIPLE SYSTEM ATROPHY

- Also see earlier section on MSA.
- Cerebellar ataxia is present in more than half of MSA patients.
- Two major types classified according to the prominent clinical presentation: Parkinsonian type (MSA-P) and cerebellar type (MSA-C)
- MSA-C starts around age 55 years with ataxia, dysarthria, and cerebellar oculomotor abnormalities.
- May also include symptoms of parkinsonism and autonomic failure.

WHEELCHAIRS

BASIC WHEELCHAIR PRESCRIPTION WRITING

- Optimal wheelchair prescription should maximize patient mobility, independence, and functional potential and minimize the risk of comorbidities and the restriction of remaining physical or neurologic limitations. There are many different wheelchairs available with numerous options from which to choose. This section covers basic principles to allow the clinician to prescribe the most appropriate wheelchair for a patient.
- Prior to writing a wheelchair prescription, the clinician should complete a full medical history, as well as assess the patient's clinical and functional status, including ROM, strength, sensation, sitting/standing balance, cognition, endurance, skin integrity, postural mat assessment, cardiovascular status, activities of daily living (ADL) performance (e.g., feeding, grooming, dressing, bathing, toileting, and transfer), instrumental ADL (e.g., child rearing, driving and

community mobility, and home establishment and management), and future rehabilitation potential. Another important consideration is to determine if the patient's status is static or progressive. Patients with a progressive condition require a wheelchair that has the flexibility to be modified as their clinical and/or functional status changes.

Basic Wheelchair Components (Figure 12–1)

- The components of a basic wheelchair are shown in **Figure 12–1**.
- In order to evaluate the particular needs of an individual for a wheelchair prescription, specific considerations and measurements based on the patient's body must be taken into account. These are typical measurements for a standard manual wheelchair **(Figure 12–2)**. For individuals who need more support or power mobility, additional measurements are necessary.

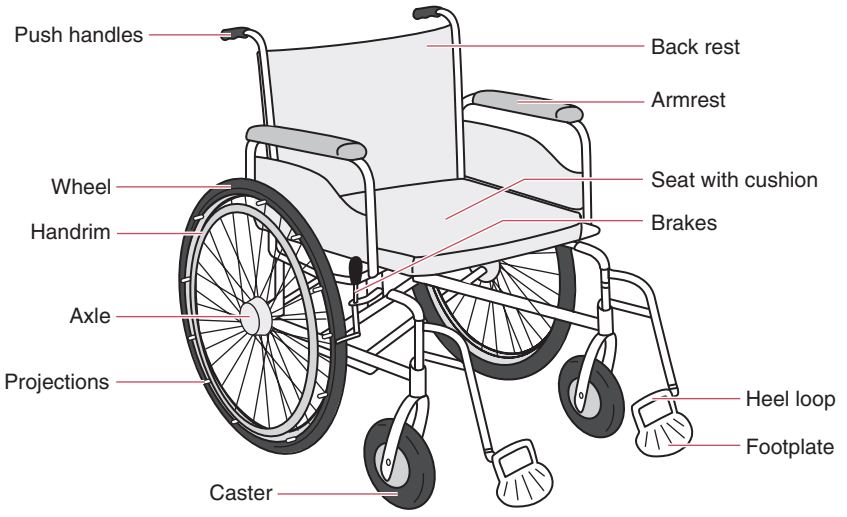


FIGURE 12–1 Manual wheelchair.
Source: Adapted from Nesathurai S. ed. *The Rehabilitation of People With Spinal Cord Injury: A House Officer's Guide*. 3rd ed. Arbuckle Publishers; 2013.

WHEELCHAIR FITTING (FIGURE 12–2)

Wheelchair Seating System Measurement

- Body measurements are typically made with the patient in the seated position because it accounts for the effect of gravity on posture. Accurate measurements are more easily taken with the patient on a flat surface, such as a treatment mat, and with the patient supported with their pelvis in as neutral a position as possible.
- 🕮 Inaccurate measurements are a major contributing factor to increased deformity, pressure injury development, and patient discomfort.

Seating Surface

- A tailored, fitted seat is important for stability, postural support, comfort, ease of propulsion, and skin integrity.
  - There are two types of seats:
    1. *Vinyl sling seat:* Easy to fold, easy to clean, and lightweight.
       - Disadvantages: The sling nature of the seat does not provide sufficient support and promotes a posterior pelvic tilt with hip internal rotation and adduction. This in turn predisposes the patient toward a collapsed trunk with corresponding head forward flexion, neck hyperextension, and a protracted and elevated shoulder girdle.

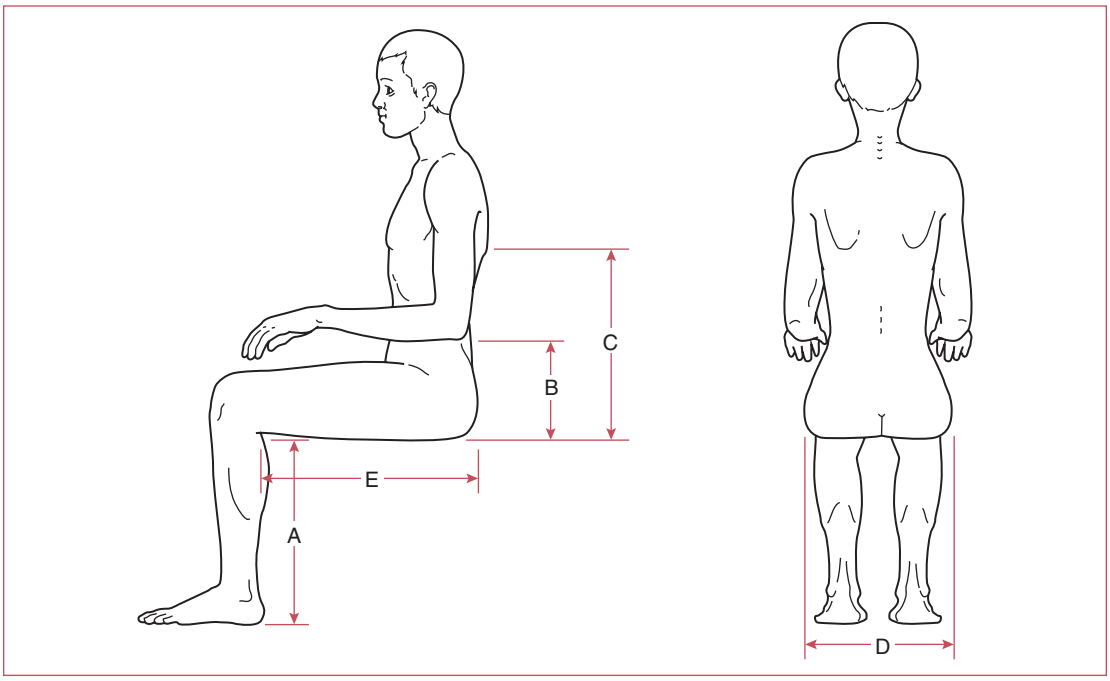


FIGURE 12–2 Critical body measurements for wheelchairs. (A) Seat height. (B) Armrest height. (C) Back support height. (D) Seat width. (E) Seat depth.
Source: Redrawn from Wilson AB Jr. *Wheelchairs: A Prescription Guide*. 2nd ed. Demos Medical Publishing; 1992, with permission.

2. *Solid seat:* Firm surface that provides a level base of support and more upright, symmetrical postural alignment.
   - Disadvantages: Slightly heavier (1–5 lb.); must be removed to fold a folding wheelchair.

- All patients should use some type of cushion. One way to provide a lighter weight yet a supportive seating surface is to place a solid seat insert under the cushion to provide a level base of support for the wheelchair seat.

Seat Width (Figure 12–2D)

- Measure is across the widest point of the hips or legs, depending on tissue distribution (with clothing and any braces or orthoses).
  - For patients who use a *manual wheelchair* as their primary means of mobility, the seat width should be the same as their hip width to maximize their wheel access. At times, for functional purposes, the wheelchair seat width may be up to 1 in. greater than the patient's measured hip width. This extra space can be beneficial for patients who perform ADLs, such as dressing or bladder management, in their wheelchair.
  - For patients who use a *power wheelchair* as their primary means of mobility, the seat width can be approximately 1 in. wider than their hip width. However, it is important to remember the risk of scoliosis can increase with pelvic malalignment. Hip guides can ensure alignment is maintained in the pelvis.
- 🕮 If the wheelchair is too narrow, transfers will be more difficult, and the patient is more likely to develop a pressure injury on the greater trochanters of the hips. Conversely, if the seat is too wide, pelvic and trunk support is compromised, which can lead to scoliosis, back pain, and difficulty with wheelchair propulsion.

Seat Depth (Figure 12–2E)

- This is determined in a sitting position, with the patient's pelvis in as neutral a position as possible and measuring from their posterior pelvis to the popliteal fossa.

- Generally, the gap between the front of the seat and the back of the knee of the wheelchair user should be 1 to 3 in. This is determined by the functional and positioning needs of the patient. One inch helps to ensure maximum distribution of the trunk weight over the buttocks and upper legs without placing undue pressure behind the knee or on the ischial tuberosities. Three inches is necessary for individuals who are expected to use their lower extremities (LEs) to propel the wheelchair or individuals who do not have grasp but will use wrist extension with a fisted hand to maneuver their leg for ADLs including bladder management.
  - If the backrest is cushioned, the thickness of the compressed cushion must be considered. If an after-market back is being utilized, it is important to measure the wheelchair seat depth from the actual position of the compressed cushion of that back support to the front of the seat sling or pan.
  - If a patient is going to be propelling the manual wheelchair with their foot/feet, 1 to 2 in. are usually subtracted, and the cushion is beveled back to allow the knee adequate flexion to propel the wheelchair.
- If the seat depth is too shallow, there is decreased weight distribution throughout the length of the femurs, resulting in increased pressure (and potential for subsequent skin breakdown) on the ischial tuberosities.

Seat Height (Figure 12–2A)

- Measure the patient's lower leg length from the bottom of the heel of the shoe to the inferior aspect of their distal posterior thigh. Subtract the height of the *compressed* seat cushion from the measurement, and then add 3 to 4 in. to allow for adequate footplate clearance.
- The measurement from the floor to the front edge of the seat will vary depending on the desired use of the wheelchair and the lower leg length of the user. A user with longer legs will require a higher seat height in order to achieve sufficient ground clearance with the footrests. The seat should be high enough to perform typical ADLs but low enough to fit under desks and tables and to efficiently propel the wheelchair.
- For the cushion height, it is important to consider the material (air, foam, or gel) and the amount of compression that occurs when the patient is seated on it. Foam cushions compress to about half their unweighted dimension. Therefore, the cushion height that is subtracted is the "compressed" cushion height.
- 🕮 Variations on seat height measurements occur in the foot drive, or hemiplegic height wheelchair, which is designed lower, with the seat closer to the floor, to allow the unaffected leg to propel the chair.
  - Measure the patient's lower leg length (as previously directed) and subtract the height of the "compressed" seat cushion only. This will allow the patient's foot to rest comfortably flat on the floor, which is essential for the patient to obtain adequate leverage and heel strike during foot propulsion.
  - If the seat height is too high for the patient, they will be constantly sliding out of the wheelchair as they attempt to obtain sufficient leverage to propel the wheelchair with their unaffected leg and arm.
  - For the patient with SCI, if the seat is too high, they may also be at risk for Achilles tendon contracture due to the ankle resting in a persistently plantarflexed position.
    - If the seat height is too low for a patient, they will be at risk for both hip flexion contracture as well as pressure injury to the ischial tuberosities.

Posterior Seat Slope

- The posterior seat slope refers to the angle of the seat with respect to horizontal. It is commonly measured in inches, with the front seat height higher than the rear seat height.
- About 1-in. posterior seat slope is common. A larger seat slope helps to increase pelvic stability by pushing the pelvis into the backrest.
- The benefit of this posterior sloped seat position is to stabilize the paralyzed pelvis to provide adequate functional reach, maximize upper limb access to the pushrim, and reduce the forces

required to propel. This wheelchair modification can reduce strain on the upper extremity joints.

- However, too large of a posterior seat slope may:
  - Increase risk of pressure injuries on the sacrum if the patient does not have sufficient hip flexion for this slope.
  - Affect the center of gravity with a decrease in the rearward stability—that is, increase the risk of tipping the wheelchair and making transferring out of the wheelchair more difficult.

Footplate Height

- Measure the distance from the heel of a patient's shoe to the posterior surface of the inferior, distal thigh at the popliteal fossa. To get the footplate height, you would then need to subtract the height of the compressed cushion and consider how much clearance from the ground the patient will need under the footrest (generally 2–3 in.).
- Footrests are usually adjustable in height and should have at least 2 in. of clearance from the floor.

Back Support Height (Figure 12–2C)

- Measure the distance from the bottom of the buttocks to the inferior angle of the scapula. Then add the "compressed" wheelchair cushion height to get the true back support height measurement.
- The height of the back support is an important measurement that affects comfort, stability, and control of the wheelchair.
  - The wheelchair back support height can vary depending on the capabilities and support needs of the patient. In general, a lower backrest height permits greater freedom of motion (e.g., leaning, turning) and is less restrictive (i.e., does not interfere with arms when pushing) but provides less support than a higher backrest. The more severely impaired an individual is, the higher the backrest must be to provide adequate postural support and trunk stability.
- For patients who will be utilizing their arms for propulsion, the height of the back support should be just below the inferior angle of the scapula. The scapula should not hang over the back support.
- If the backrest is too high, it may block the scapula and interfere with shoulder movement for optimal joint protection with propulsion. If it is too low, it will not provide adequate trunk stability.
- If the patient has good trunk control and can propel a wheelchair, the back support can be lower than just under the inferior angle of the scapula. The only caution is to make sure that the patient is not hyperextending their back over the back support for stability.
- If the patient has no upper extremity strength and poor trunk control, the measurement is first taken by measuring the patient from the bottom of the buttocks to the spine of the scapula. Again, the compressed wheelchair cushion height is added to this measurement to get the back support height for a patient with poor upper extremity and trunk control. Typically, the back support does not need to be higher than the patient's shoulder and should be just under their shoulder at the spine of the scapula. This patient will also need to have a headrest for adequate support.
- If a patient requires an anterior chest harness, which is more common in the pediatric population, the back support height needs to be to the top of their shoulder so that the harness is supporting them optimally and is not compromising their posture.

Head Support

- Measure from the bottom of the buttocks to the top of the cranium. Then add the height of the "compressed" seat cushion to determine the "top of the headrest" position. This is adjusted down by 2 to 4 in. depending on the patient's support needs and/or preference.

Armrests (Figure 12–2B)

- Measure arm height from the buttocks to the bottom of the patient's bent elbow at 90 degrees. Then add the height of the compressed seat cushion to obtain the armrest height.
- Wheelchair armrests may be fixed or removable (swing-away, flip-back style, or fully removable); adjustable or fixed height; full length or desk length; and contoured with foam over the tubular material or standard flat pad over the tubular material.
  - Wheelchair armrests are essential to provide appropriate glenohumeral support. They provide patients with a surface to shift their weight onto when reaching and functioning from a wheelchair, as well as a surface for patients who must elevate their body at regular intervals to perform a "push-up" to relieve pressure and minimize their risk of pressure injuries.
  - If armrests are too high, it will lead to poor posture and shoulder discomfort, and the wheelchair will not fit under the table. If they are too low, it could lead to shoulder discomfort, poor posture, and compromised respiration.

FIXED VERSUS REMOVABLE FLIP-BACK ARMRESTS

- *Fixed armrests* are lighter because of fewer moving parts and do not add width to the chair.
  - Disadvantages: They are not usually prescribed, as they do not allow a patient to perform lateral transfers to/from the wheelchair.
- *Removable, swing-away, or flip-back style armrests* are essential for patients who need to perform lateral transfers to/from the wheelchair.
  - Disadvantages: Added weight to the wheelchair. Some removable armrests also increase the overall width of the wheelchair by 2 in. However, if it is flip-back style, one can prescribe a wraparound armrest to minimize this (see below).

FULL-LENGTH VERSUS DESK-LENGTH ARMRESTS

- 🕮 Fixed and removable styles are available. The removable desk-length armrest is most commonly prescribed.
- Full-length armrests are approximately 14-in. long and offer more arm support, which is essential to support a lap tray or arm trough. They are also necessary for upper extremity support with sit-to-stand transfers to/from the wheelchair.
- The disadvantage is that the patient will be unable to get close to tables and desks.
- Desk-length armrests are approximately 10 in. and provide the patient with better access to tables for feeding, sinks for grooming, and desks for working.

ADJUSTABLE HEIGHT ARMRESTS

- Adjustable height is important to provide each patient with adequate and specific glenohumeral support. It is available as an alternative to ordering a fixed height.
- The armrest height is dependent upon the patient's height and body type (short or long limbed), as well as the cushion used and its compression. This results in a wide range of measurement that is very different with each patient.
  - Disadvantage: Adjustable armrests are heavier than fixed.

TUBULAR VERSUS STANDARD ARMRESTS

- Tubular armrests allow the patient to obtain better forearm clearance when propelling the wheelchair. The square edge of the standard armrest pad often causes forearm bruising with propulsion.
- Tubular armrests are more lightweight than the standard removable or flip-back armrests.

OTHER ARMREST VARIATIONS

- Depending on the manufacturer, flip-back armrests can come in a "wraparound design" where the armrests attach behind the seat. This design feature minimizes the increase to the overall

width of the wheelchair. This is commonly ordered to maximize a patient's support as well as environmental access.
- Swing-away or flip-back armrests are preferred by active patients with SCI to enable them to transfer independently.
- Active, younger patients often prefer no armrests if balance is not a concern.

Recline and Tilt-in-Space Mechanisms

- The most important reasons for a tilt-in-space and/or recline seating system is to provide a patient or caregiver with the ability to perform a position change. This is essential for an adequate weight shift for pressure redistribution for the patient's body to minimize the risk of pressure injury and to provide the patient with postural support from gravity for upright sitting. Other indications for use may include poor sitting balance, poor endurance, orthostasis, fluctuating respiratory status needs, back pain, and the need to be able to adjust the backrest for ADLs.
- These systems can be used in manual or power wheelchairs.
- It is important to educate the patient and caregiver(s) about these systems, as they add size, weight, and bulk to the wheelchair. In addition, they generally require a longer wheel base to maintain adequate stability when the chair is reclined or tilted. These are important considerations for home accessibility. Many of the tilt-in-space systems now come with a center of gravity-type mechanism where the seating system shifts forward on the wheel base as it tilts to minimize this overall length increase.

RECLINER WHEELCHAIR

- A reclining back support can be easily repositioned incrementally in a posterior direction from an upright to various reclined positions. The seat of the wheelchair stays in the same position relative to the floor while the back support is adjustable. A recliner can be a semi-recliner that stops at 120 degrees or a full recliner that stops at 180 degrees.
- A manual wheelchair with a reclining back is at minimum 3 in. longer than a standard wheelchair and is more difficult for a patient to propel because of the additional weight and a more posterior wheel base position. Also, the more a chair can recline, the longer the overall wheelchair will be.
- It is important to remember that shear forces are increased as an individual is being reclined and unreclined in the wheelchair.
- In power wheelchairs, there are recliner seating systems with reduced shear, known as low and zero shear recliners. However, no system will completely eliminate shear forces. This is an important consideration for patients with prominent vertebral processes or patients with more bony body frames.
- *Recliner advantages:*
  - Independent pressure redistribution
  - Postural support from gravity
  - Pain relief
  - Assistance in orthostatic episodes
  - Allows for PROM of hips—however, it is not a substitute for a PROM program
  - Easier to perform bladder catheterization
  - Can help with dressing/undressing
  - Can help mobilize secretions
  - Can maintain items on lap tray during a weight shift
  - Can maintain position under table or desk during "pressure relief"
  - If elevating leg rests are included with this recline, results in optimal pressure redistribution throughout the entire posterior of a patient's body
  - In a manual version, a reclining wheelchair can be transported in a car with a large trunk.
- *Recliner disadvantages:*
  - Increased shear forces can result in shearing pressure injuries

- Can increase spasticity due to the hip position change
- Less maneuverability due to increased turning radius
- Patient does not maintain an "ideal" seated position after several weight shifts

TILT-IN-SPACE SYSTEM (FIGURE 12–3)

- Alternative wheelchair system that can also provide posterior body position for pressure redistribution or postural support.
- 🕮 The angle of the seat and back does not change when the seating system is tilted back. The patient remains in the same position, but orientation in space changes. This allows pressure to be redistributed from the seating surface to the back support. Since there is no movement of the back away from the seat, shear forces are minimized.
- 🕮 In order to redistribute sufficient pressure for a weight shift, a tilt-in-space system alone must tilt at least 45 degrees. Most patients need more than 45 degrees, and many systems provide a range of 45 to 60 degrees of posterior tilt.

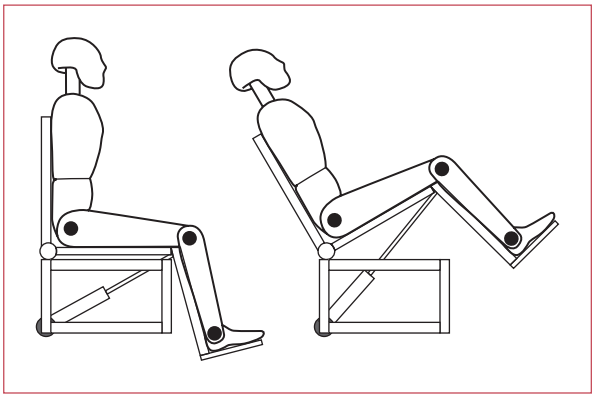


FIGURE 12–3 Schematic for tilt-in-space mechanism. The user remains in the same position while the orientation changes. This allows pressure to be redistributed.
Source: Redrawn from Cooper RA. *Wheelchair Selection and Configuration*. Demos Medical Publishing; 1998, with permission.

- 🕮 *Tilt-in-space* advantages:
  - Pressure relief
  - Postural support from gravity
  - Pain relief
  - Can assist in orthostatic episodes
  - Minimizes shear
  - Diminishes effects of spasticity during position changes
  - Maintains seating position during weight shifts
  - Helps mobilize secretions
  - Smaller turning radius; therefore, more maneuverability than a reclining wheelchair
- *Tilt-in-space disadvantages:*
  - No PROM benefits
  - May not offer as much pressure distribution relief as a recliner to 180 degrees
  - If using a leg bag, urine may run backward in the tilted position. An antireflux valve on leg bag tubing can manage this.
  - Difficult to maintain items on a lap tray when tilted
  - Need to come away from desk or table to perform position change
  - More difficult to perform bladder catheterization
  - In a manual wheelchair, most of these systems do not disassemble sufficiently for transport by car.

RECLINE VERSUS TILT-IN-SPACE SEATING SYSTEMS

- The previous comparisons between tilt-in-space and recliner wheelchairs were made to assist the patient in deciding on which product may best meet a patient's medical, physical, functional, and lifestyle needs.
- Despite the number of advantages versus disadvantages, either of these systems could be the "perfect" wheelchair for a particular patient. It is optimal for many patients to have both tilt-in-space and recline seating systems together in the same wheelchair to maximize their overall functioning at a wheelchair level and their level of pressure redistribution for effective pressure relief.

- The duration of pressure redistribution with 35 degrees of tilt and 120 degrees of recline is recommended for 3 minutes for adequate perfusion of blood flow to the ischial tuberosity region back to baseline level in individuals with SCI.
- A combination of tilt and recline is more commonly recommended in power wheelchairs over manual wheelchairs due to the weight of the components for these seating functions.

Wheels (Figure 12–4)

- The standard chair comes with 3- to 8-in. diameter front caster wheels and 24- to 26-in. diameter rear wheels.
  - Mag wheels are most common in standard wheelchairs. They are one-piece cast with metal alloys or metal and plastic to weigh only slightly more than the wire spoke wheel, and maintenance-free.
- Spoke wheels are similar in design to a bicycle wheel and are lighter weight, which translates into less force needed for propulsion. They also have improved shock absorption but require more maintenance secondary to bending and loosening. Annual maintenance includes tightening the spokes.
- There are wheels that are lighter weight and more durable than standard spoke wheels. These include Spinergy LX, Spox®, and Carbon Blade®. They are more durable and require less maintenance than a standard spoke wheel but are also more expensive.

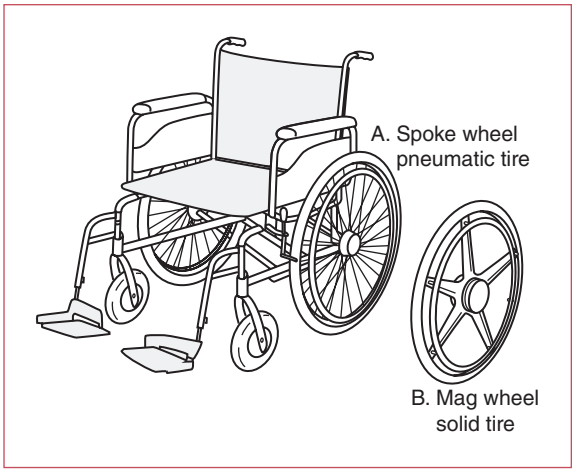


FIGURE 12–4 Wheels for wheelchairs. (A) Spoke wheel, pneumatic tire. (B) Mag wheel, solid tire.
Source: Redrawn from Wilson AB Jr. *Wheelchairs: A Prescription Guide*. 2nd ed. Demos Medical Publishing; 1992, with permission.

WHEEL AXLE POSITION (FIGURE 12–5)

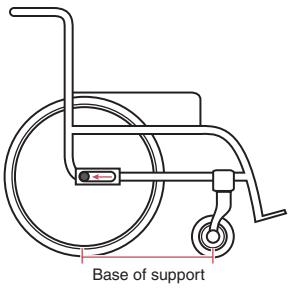
FIXED AXLE

- Most standard wheelchairs have a fixed axle position. It is the one location where the wheel attaches to the frame. In a standard wheelchair, this is positioned toward the rear of the wheelchair and is appropriate for someone who is not using the wheelchair as a primary means of mobility.
- In an ultralight wheelchair, this is a highly customized option that is ideally configured with the patient seated in the wheelchair in the exact seating system they are looking to use in the wheelchair.

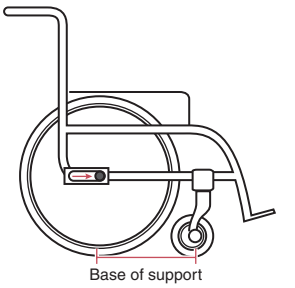
ADJUSTABLE AXLE

- 🕮 In a wheelchair with an adjustable axle, the wheel axle can be moved forward to position the center of gravity underneath the patient to allow for improved rear wheel access, increased upper limb joint protection, and more efficient wheelchair propulsion. In addition, this forward axle position facilitates "wheelies." This is important for function in the patient with SCI. "Wheelies" are an essential basic wheelchair skill that is necessary to safely transition up/down curbs.
- 🕮 The axle can also be moved posteriorly using a special plate or adjustable frame design to increase wheelchair stability. This is to compensate for the change in the location of the center of gravity, such as in the absence of legs in a bilateral amputee patient, and is often a standard starting position with a reclining back or tilt-in-space wheelchair.
- 🕮 The more posterior the rear wheels:
  - The greater the rolling resistance
  - The more energy required for propulsion

  - The greater the turning radius
  - The more stable the chair
  - The more extension needed at the glenohumeral joint, which can increase repetitive stress injuries at the shoulder
- 🕮 The more anterior the rear wheels:
  - The less the rolling resistance
  - The less energy required to propel
  - The smaller the turning radius
  - The less stable the chair, the more maneuverable the chair (easier to perform a "wheelie").
  - Improved upper limb wheel access for more efficient propulsion
  - May relieve shoulder pain during manual propulsion



Posterior wheel position increases stability



Anterior wheel position increases maneuverability

FIGURE 12–5 Adjusting axle position. *Source:* Adapted from Nesathurai S, ed. *The Rehabilitation of People With Spinal Cord Injury: A House Officer's Guide*. 3rd ed. Arbuckle Publishers; 2013.

Tires

- There are several types of tires available for use with different types of wheel options. For individuals with SCI, who often use an ultralight wheelchair as their primary means of mobility, pneumatic tires would provide the lightest and smoothest ride. However, the risk of a flat tire often outweighs the benefits for many patients.
- ***Solid rubber tires:***
  - Have a very low rolling resistance on flat or smooth surfaces
  - No risk for "flat" tire
  - Lack "cushioning" on rougher terrain; rougher ride than a pneumatic tire
  - Heavier than a pneumatic tire
- ***Pneumatic tires:***
  - They are lighter than nonair options.
  - They provide a smoother, more comfortable ride than solid tires.
  - The higher the pressure square inch, the lower the rolling resistance.
  - They often contain an air-filled inner tube and are lightweight.
  - The profile of the tire can be highly textured or smooth.
  - The best ride on most surfaces, but not as good as rubber on smooth surfaces.
  - 🕮 If there is carpeting on most surfaces, pneumatic tires are best because carpeting increases the rolling resistance by a factor of 4. Pneumatic tires also provide cushioning for outdoor use to allow a more comfortable ride; they also reduce wheelchair wear and tear.
  - Maintenance consists of replacing air in the tires every 3 weeks.
  - Can run the risk of a "flat" tire.
  - Pneumatic tires also come with an airless (flat-free) insert that is a soft rubber or latex gel that replaces the inner tube. The ride is cushioned and it does not go flat. However, the airless insert is as heavy as a solid tire.
- ***All-terrain tires***
  - They have a wider tread and are wider overall.
  - They are pneumatic tires that require a standard inner tube.
  - They are used for mobility on soft and sandy terrain and are up to 2.5 in. thick.
  - For smooth surfaces, there is a lot of rolling resistance for the user.
- ***Kevlar® tires:***
  - They are made of Kevlar, which is more puncture-resistant.
  - They provide a durable and smooth ride.
  - Lower risk of a "flat" tire compared with a standard air tire.

CAMBER

- **Camber** is the wheel angle against the vertical axis where the bottom of the wheel is out further than the top of the wheel.
- Camber positions the wheel in a better biomechanical position to the shoulder joint.
- Camber has several advantages:
  - Camber maximizes lateral stability of the wheelchair because it widens the footprint of the chair creating greater side-to-side stability.
  - Low degrees of camber allow for easier propulsion of the wheelchair (especially at higher speeds), as it positions the pushrims more ergonomically for propulsion (it is more natural to push down and out).
  - Camber also helps to protect the hands because the bottom of the wheels will scuff the edges of obstacles, which prevents the user's hands from hitting the obstacle and makes it easier to turn.
- The disadvantages are the increased overall width of the chair, making it difficult to maneuver indoors, and increased tire wear on the inside part of the tire. This can be managed by choosing smaller degrees of negative camber angle and making sure the overall width is less than a standard doorway (<28 in.).
- Camber is available in increments ranging from 2 to 12 degrees. For an everyday wheelchair, most patients function with 2 to 5 degrees of camber. The width of the wheelchair seat often dictates the degree of camber that is chosen to ensure that the wheelchair can still fit through most doorways.
- Camber angle can either be fixed or adjustable, depending upon the frame design. The width of the chair depends upon the width of the frame, the handrim position, and the camber angle.

HANDRIMS

- Handrims are round or oval surfaces attached to the large rear driving wheels to provide the patient with a place to propel the wheel without touching the tire directly to minimize soiling the hands.
- Handrims are also smaller than the wheel, making the chair easier to propel.
- The larger the diameter of the handrim, the easier it is to grasp and propel. The concern with this is the increased weight.
- The *standard handrim* is a circular aluminum or steel tube. However, for individuals who have difficulty with gripping a smooth surface, vinyl, foam, rubber, or plastic coating is available with optional glove use to provide this patient's hand with increased traction.
- Another option is the *"natural fit handrim."* This is an oval-shaped handrim with or without a thumb guard that promotes a more ergonomic wrist and hand position on the handrim. While this handrim is heavier, most patients find that the benefits of this more ergonomic handrim outweigh the increased weight concern.
- There are also vertical, horizontal, or oblique projections to improve propulsion. However, it is important to remember that horizontal and oblique projections increase the overall width of the chair.
- For individuals with a brachial plexus injury, upper extremity amputation, or hemiplegia **(Figure 12–6)**, there is an option to obtain both handrims

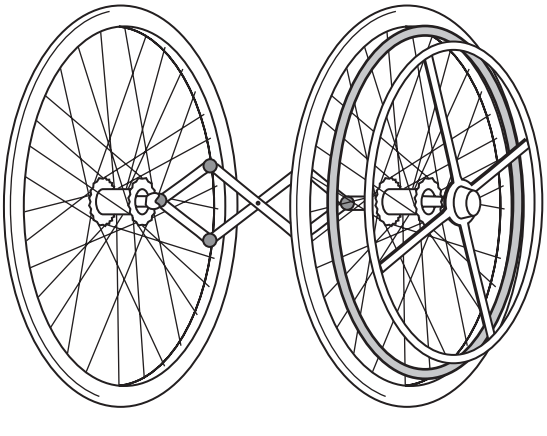


FIGURE 12–6 Mechanism for a one-arm drive wheelchair. *Source:* Redrawn from Wilson AB Jr. *Wheelchairs: A Prescription Guide*. 2nd ed. Demos Medical Publishing; 1992, with permission.

positioned on one side of the wheelchair wheel. This is called a one-hand-drive wheelchair. The wheelchair has interconnected driving wheels so that both wheels can be controlled from one side through a dual set of handrims. When one handrim is moved independently of the other, only one wheel moves. When both rims are moved together, both wheels are driven simultaneously. The major concern is the increased potential of a repetitive stress injury from the patient's one functioning upper extremity performing a repetitive task. A power wheelchair is often the best option for this patient to minimize the risk for increased injury and disability.

CASTERS

- Casters are the small front wheels that allow turning of the wheelchair and are available in 2-, 3-, 4-, 5-, 6-, and 8-in. sizes.
- The smaller and narrower the caster, the lighter and more maneuverable the chair.
- 🕮 A smaller caster provides a smaller turning radius, but if it is too small, it can perform poorly on outdoor surfaces and on carpets because it can get stuck in cracks on sidewalks and in uneven contours of an uneven surface.
  - Many patients obtain tremendous benefit from increased maneuverability.
- Larger diameter casters make it easier to negotiate uneven surfaces but have larger turning radii and therefore offer less maneuverability in normal tight indoor environments.
- The 8-in. diameter caster is standard on the basic wheelchair. The larger size is great for uneven indoor and outdoor surfaces, such as carpet, sidewalks, and grass.
  - The disadvantage of an 8-in. caster is the decreased maneuverability and increased strain on the upper extremity to perform frequent small adjustments to maneuver in tight areas indoors, such as within bathrooms, or to maneuver at a table or desk.
  - 8-in. casters may also be ordered with pneumatic or semipneumatic tires to be used on rough surfaces or outdoors.
- The 5- and 6-in. caster sizes tend to be the average caster size that gives a patient a good balance of indoor maneuverability and good outdoor performance.
  - The 5- and 6-in. caster sizes are used in many ultralight chairs and on children's chairs.
- Casters can be a-solid polyurethane type, semipneumatic, or "soft roll." In addition, the 6- and 8-in. casters can be pneumatic. The advantage to pneumatic is a softer ride, and the disadvantage to pneumatic is the more difficult maintenance with this small front wheel and the risk of a "flat" tire.
- 🕮 Moving the caster placement more posterior decreases the turning radius, increases maneuverability, but decreases anterior stability. When this is performed, it is important to test with the user for sufficient anterior stability.

FRONT RIGGING

- Front rigging is the term used to describe the footrest and leg rest collectively.
- A footrest consists of a support bracket with a footplate and must provide sufficient support to hold the feet in proper position to prevent foot drop or other deformities.
- 🕮 Footrests are measured by taking the distance from the heel to the under surface of the thigh at the popliteal fossa. The "compressed" cushion height should be accounted for. Footrests are usually adjustable with a wrench and should have 2 in. of clearance from the floor.
- *Footrests* may be fixed, folding, or swing-away.
  - *Swing-away footrests* are the most common and allow for safer transfers, as they swing out of the way of the feet. Swing-away removable footrests help with portability of the chair. A swing-away footrest is heavier than a fixed footrest due to the additional components needed to remove them. They should include a heel loop to stabilize a patient's foot on the footplate.
  - *A fixed footrest* is often tubular and come with or without a footplate. They are often utilized to maintain a patient's weak or paralyzed LEs on the wheelchair. Fixed footrests allow for a lighter chair but are a safety concern for individuals who perform sit-to-stand transfers.

A fixed footrest is more common for a patient who is not standing and is performing lateral transfers to get into and out of their wheelchair.

- An *elevating leg rest* consists of an elevating support bracket, a footplate, and a calf pad to support the back of the leg when elevated. In a manual wheelchair, elevating leg rests typically have a swing-away mechanism.
  - Elevating leg rests help with decreasing dependent edema and come with a calf support for the lower leg.
  - Elevating leg rests are necessary for patients with limited knee flexion, including severe knee arthritis, below-the-knee amputation, knee-extension contractures, or other joint abnormalities.
  - Elevating leg rests should not be used for patients who have tight hamstring musculature.
  - Three major disadvantages to an elevating leg rest on a manual wheelchair:
    - Significantly increased weight of the leg rest (compared with a standard swing-away footrest).
    - Elevating mechanisms require frequent repair.
    - Increased overall length of the wheelchair.
  - If a patient does not have orthopedic knee limitations, they can elevate the LEs on a footstool or chair to minimize the concerns with increased weight, maintenance, and decreased maneuverability.
- Footrests and leg rests may be ordered differently for each LE depending on a patient's particular needs. For configuration to a patient's body:
  - A footrest that is too low (too long) will increase pressure over the lower posterior thigh and/or hit the floor on uneven surfaces.
  - Conversely, a footrest that is too high (too short) places the knee higher than the seat and shifts the patient's weight posteriorly, which increases pressure over the ischial tuberosities and sacrum. This can increase the risk of pressure injury to these regions.

Wheelchair Weights

- *Standard*—usually steel construction, 40 to 65 lb.
- *Lightweight*—usually aluminum or steel material, 30 to 35 lb.
- *Ultralightweight*—usually aircraft quality aluminum, <30 lb.
- *Titanium and carbon fiber*—<20 lb.

Wheelchair Cushions (Figure 12–7)

- Many different designs of seat cushions are available. Selection of seat cushions may be divided into the five basic types shown in **Figure 12–7**. In addition to these, there are also many combinations of these materials. This chart provides an introduction to obtain a good initial understanding of the pros/cons of the various materials. All wheelchairs should be used with a seat cushion. Patients should not sit directly on the constructed seat (sling or rigid) of the wheelchair.
- Seating should provide good pressure distribution with appropriate LE and pelvic alignment, enhance trunk and pelvic stability, and provide comfort. Cushions should be durable. The most appropriate medium should be chosen to maximize postural and LE support and to provide an optimal microclimate for maximum skin protection. If incontinence is a concern, an incontinence cover can help. If significant perspiration is a concern, a medium with more air exchange (air vs. foam) should be considered.

Safety Equipment

- When writing a wheelchair prescription, it is important to include basic safety equipment.

PELVIC BELT

- Pelvic belts should be worn for safety and to maintain pelvic alignment while sitting in the wheelchair. The patient may be extremely cautious while seated in the chair, but unforeseen circumstances surrounding the individual may jar the chair enough to send a patient out of the chair.

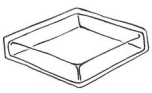
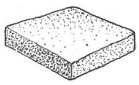
					
CUSHION MATERIALS	**VISCOUS FLUID**	**AIR**	**GEL**	**VISCOELASTIC FOAM**	**FOAM**
Envelopment (ability of the cushion to surround the pelvis)	Variable	Variable	Poor	Good	Good
Shear (forces that occur when adjacent surfaces slide across one another)	Low	Variable (dependent on cover)	Low/medium	High	High
Thermal properties (ability to transfer heat and cold)	Good	Variable (dependent on cover)	Good	Good	Poor
Damping (ability to soften impact)	Poor	Good	Poor	Poor	Good
Weight	Heavy	Lightweight	Heavy	Lightweight	Lightweight
Maintenance	Minimal	Significant	Minimal	Minimal	Minimal

FIGURE 12–7 Wheelchair cushions.
Source: Adapted from Lange, M. Cushion comparison. 2012. http://physical-therapy.advanceweb.com/Features/Articles/Cushion-Comparison.aspx.

WHEEL LOCKS (FIGURE 12–8)

- Wheel locks, also known as brakes, secure the rear wheels of the wheelchair to avoid rolling away on uneven surfaces and to provide stability during transfers. Wheel locks should not be used to slow a chair down. An abrupt stop can result in overturning the wheelchair.
- The basic types of wheel locks available include push-to-lock lever, pull-to-lock lever, and scissor.
- Selection is based on available upper extremity and hand function. Wheel locks may push or pull closed and can be mounted low or high. With swing-away footrests and leg rests, it is recommended to use pull-to-lock wheel locks so that the wheel lock does not become disengaged when the footrest/leg rest is swung out to the side for lateral transfers.
- Low mount and scissor type wheel locks are helpful to position the wheel lock out of the way during normal wheel propulsion. This minimizes the risk of a patient hitting their thumb on the wheel lock during wheelchair propulsion. Good balance is needed for the patient to reach the low mount and scissor locks.

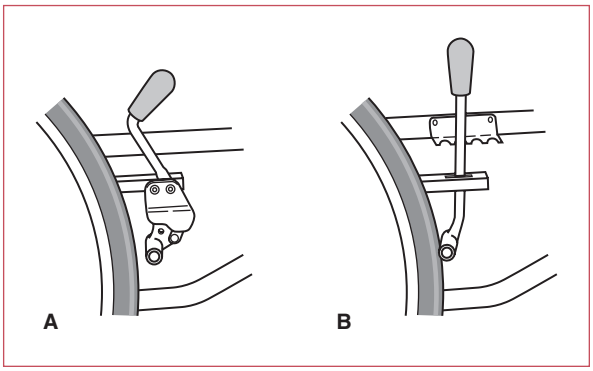


FIGURE 12–8 A push to lock wheel lock: (A) toggle type; (B) lever type. Variations of these two types of brakes are available.

- High-mounted locks are easier to negotiate but may interfere with transfers.
- Wheel lock extensions can be placed over the lever wheel locks for patients who need more leverage to access the wheel locks and for patients who cannot reach from the contralateral side, such as a patient with hemiplegia from a stroke.

GRADE AIDS

- Grade aids prevent the chair from rolling backwards on an incline. This is necessary and helpful for patients with limited strength and endurance, particularly where inclines prove challenging due to steepness or length.

ANTI-TIPPERS

- Anti-tippers can be fixed or removable and are extensions most commonly added to the lower and posterior frame of the wheelchair to minimize the risk of the wheelchair tipping backward. They are also available for attachment to the front rigging to avoid forward tipping in unique situations.
- Anti-tippers are typically used on recliner wheelchairs, for patients who are unable to "pop" a wheelie independently, who have poor to fair control with stand-to-sit transfers, above-the-knee amputations, ataxia, or SCI.
- It is important to note that anti-tipping devices may interfere with curb and step negotiation. They are typically designed so that they can be rotated upward for clearance to allow for safe negotiation of steps and curbs for functional wheelchair mobility.

WHEELCHAIR STABILITY

- Wheelchair center of gravity is affected by the user's posture, weight, and presence of limb amputations.
- Wheelchair stability can be impaired by the essential addition of equipment for life support, communication aids, and assistive technology, and should be tested with the patient and all of their essential equipment.
- Assessment of wheelchair stability can help guide modifications to decrease falls.

INTELLIGENT WHEELCHAIR SYSTEM

- The intelligent wheelchair system assists persons with cognitive impairments to more safely operate a powered wheelchair.
- Through a visual-based anticollision component, the intelligent wheelchair system prevents the wheelchair from colliding with obstacles. Additionally, the navigation assistance component provides audio prompts to maneuver around potential obstacles.

Pushrim-Activated Power-Assist Wheels

- Pushrim-activated power-assist wheels (PAPAW) can be added to a manual wheelchair, allowing the wheelchair to go farther distances from pressure applied to the pushrims.
- Based on the model, power assistance can be proportional to the patient's effort or can be preset ahead of time independent of effort once the patient triggers power assist.
- Some examples include the Alber e-motion and Twion wheels (Invacare), the Xtender (Sunrise Medical).
- Advantages:
  - Decreased stroke force required for propulsion
  - Reduced electromyographic activity of muscles involved in propulsion
  - Lower oxygen consumption (energy demand) and heart rate when compared with a manual wheelchair at different speeds
  - Decreased heart rate on outdoor course when compared with manual wheelchair

  - Power assistance may reduce the risk of upper limb injury, neuropathy, and fatigue in the manual wheelchair user population.
- Disadvantages: Assistance required to load/unload wheelchair in/out of a car when compared with an ultralight wheelchair without this component.

Power-Assist Devices

- The Smart Drive (Max Mobility) power-assist wheel can be added to a manual wheelchair and will activate differently from the pushrim-activated wheels above. The Smart Drive MX2+ has a power wheel mounted on the rear of the wheelchair and the patient will have a wristband (an Apple watch or Google Ticwatch with the Push Tracker E2 software) on that communicates via Bluetooth to the wheel. The patient taps their arm with the wristband twice to turn on the wheel, then taps once to set the speed as it accelerates up. This will allow the wheelchair to keep going at a desired speed from the patient. The Smart Drive will stop when the user taps the arm with the wristband twice. At this time, the user stops the wheelchair with the handrim on the wheelchair.
- Advantages:
  - Decreased stroke frequency required for propulsion
  - Lower oxygen consumption (energy demand) and heart rate when compared with a manual wheelchair at different speeds
  - Power assistance may reduce the risk of upper limb injury and neuropathy in the manual wheelchair user population.

Power Add-On Devices

- If an individual requires a joystick to mobilize themselves but does not have the capacity to integrate a power wheelchair into their lifestyle (e.g., transportation via family car vs. a van or several steps to enter/exit home), there are various power add-on devices that can turn a manual wheelchair into a power wheelchair. That said, it is important to remember that even the most robust, ultralight manual wheelchairs were not manufactured to withstand the forces that it will sustain with a power add-on product. As a result, there may be more frequent maintenance necessary to keep this type of wheelchair in optimal working condition.
- It is essential that an individual in this type of product can perform an independent weight shift to minimize their risk of pressure injuries with a typical functional day.
- The Invacare Alber Efix and the Spinergy ZX-1 are two examples of a power add-on device.

POWER WHEELCHAIRS

- A power wheelchair can be classified in many different ways. The easiest way to understand power wheelchair technology is to break it down into three components: the power wheelchair base, the seating system, and the control system.

Wheelchair Base

- Most models can be classified as either a rear wheel drive, mid-wheel drive, or front wheel drive base, based upon the position of the drive wheel (the largest wheel). **Figure 12–9** shows a typical rear wheel drive base power wheelchair because the larger drive wheel is at the rear of the wheelchair. There is not an ideal drive base, as all of them have advantages and disadvantages.
  1. Rear wheel drive:
     - Maneuvers similar to a manual wheelchair (large wheel in the rear) and is typically the easiest model to drive as the user has the most amount of visual information from the environment in front of them with the large wheel in the rear.
     - There may be less maneuverability due to a longer overall length or width of the chair based upon the placement of the footplates relative to the front casters.

- As the weight of the wheelchair is in the rear, this model is more stable with descending hills, and conversely if a user goes up a steeper hill, these wheelchairs are not as stable as the other models posteriorly. To control for this, they are designed with a pair of suspended anti-tipper wheels in the rear.

2. Mid-wheel drive:
   - Has the larger drive wheel in the middle of the base. Due to this design, it requires front and rear wheels to keep the wheelchair stable in both anterior and posterior directions.
   - Relatively easy to drive because the center of mass (the person's body) is over the large drive wheel. Consequently, it can turn on its own axis, similar to a pirouette. This translates to a wheel base that has the smallest overall turning radius for enhanced maneuverability in tight spaces.
   - At higher speeds, the user may not feel as "in control" as they do with a rear wheel drive wheelchair.
   - The rear wheels are maneuvering posterior to the user's body and therefore is out of their visual field. If the user does not have a good perception of the rear of the wheelchair, they may hit into objects unknowingly.
   - Depending on the suspension in the wheel base and the amount of unevenness with the environment (a steeper curb cut, door saddle, or very uneven grass), the rear wheels and the front wheels on a higher surface will result in suspending the middle drive wheel (known as "high centering") and becoming stuck in that uneven environment.
3. Front wheel drive:
   - Has the drive wheel in the front of the wheelchair, resulting in a larger front wheel to transition over obstacles more easily. The pair of suspended anti-tipper wheels is in the front.
   - A major advantage is how close the front of the wheelchair is to their environment and the smallest "front turning radius" of the three models.
   - From a driving perspective, the turning with this wheelchair occurs behind the user's body. The user therefore needs to have a good understanding of where the rear of the wheelchair is, and as such requires a higher level of skill to drive than the other two drive bases.
   - With the weight of the wheelchair in the front, this model is more stable with ascending a steeper hill and less stable with descending a hill.

Seating System

- There are three different levels of seating:
  1. A basic "sit and go" seating: Similar to a car seat.
  2. Rehabilitation seating: A plethora of seat cushion and back support options can be used on the wheelchair to better meet a user's postural support and skin integrity needs.
  3. Power seating system: This is the highest level and can be composed of power tilt, power recline, power leg rests, a power seat elevator, and a power drop-down footplate. Some models even have a power standing system.

Electronics Capability

- There are three levels:
  1. Consumer power: Typically, a joystick is used to maneuver the wheelchair, and there is minimal to no programmability to the electronics. This is best for an individual with good upper extremity ROM and control but may have an endurance or orthopedic issue, where they would benefit from power mobility.
  2. Programmable electronics: Typically, a joystick is used to maneuver the wheelchair, and the electronics can be configured to better match an individual's drive controller needs. For example, an individual with a neurologic injury or coordination issue that requires a change to acceleration, deceleration, or turning for maximum control and safety with mobility using the joystick.

3. Programmable electronics that allow for different drive controllers such as a sip and puff system or a head array drive control system. There are many variables (speeds, acceleration, deceleration/braking) that can be adjusted to maximize an individual's ability to use the joystick safely.

Control Mechanisms for Power Wheelchairs (Figure 12–9)

JOYSTICK CONTROL

- The patient uses their hand to move a joystick for wheelchair control. The joystick acts as both the steering wheel and the gas pedal. It maneuvers the wheelchair in the direction it is pointed. The harder the control is pushed, the faster it will go. Joysticks mounted for hand control are available to be mounted with an array of different hardware and with a plethora of different handles in a variety of sizes, shapes, and configurations to accommodate strength, dexterity, and ROM deficits.

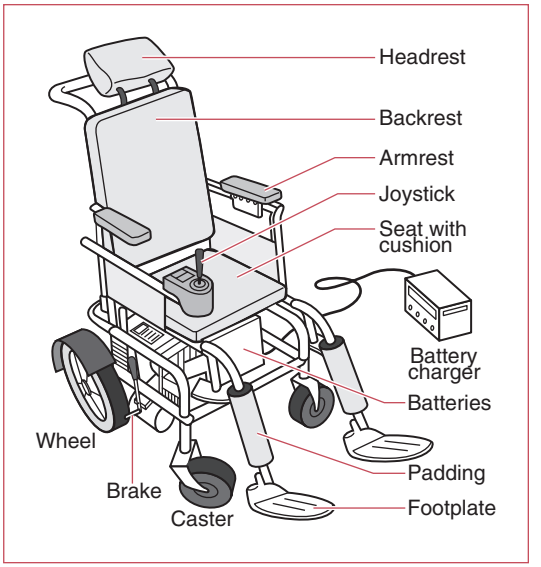


FIGURE 12–9 Typical power wheelchair. *Source:* Adapted from Nesathurai S, ed. *The Rehabilitation of People With Spinal Cord Injury: A House Officer's Guide*. 3rd ed. Arbuckle Publishers; 2013.

HEAD CONTROL

- A specialized headrest with proximity switches provides the patient with the ability to use their head to control wheelchair driving and seat function operation. With higher level electronics, a patient may also be able to access certain items in their environment, such as phone, tablet, home lights, or the computer mouse. A head array is often utilized with an emergency or reset switch to maximize a patient's safety and efficiency with operating the different modes: Mobility, seating system for position change, or phone for communication.

CHIN CONTROL

- A standard or mini joystick is mounted on either a specialized chest support or a headrest or back support mounted bracket. The chin control is positioned directly in front of the patient's chin. This joystick operates the same as the hand-mounted joystick described previously.

PNEUMATIC CONTROL ("SIP AND PUFF" CONTROL)

- The patient uses air from the mouth to activate the wheelchair through a straw.
- The hard puff is typically a forward command, a soft puff is a right command, a soft sip is a left command, and a hard sip is a stop. Once the wheelchair is stopped, the soft sip can also be a reverse command.
- Pneumatic controls are also typically utilized in conjunction with a reset switch, which provides a patient with the ability to access the wheelchair electronic functions for drive control, power seat function access, and environmental control as described with the head array using breath through the sip and puff straw.
- Finally, a reset switch can also be utilized as an emergency stop switch in the event that the patient loses contact with the sip and puff straw and cannot provide the necessary hard sip to stop the wheelchair (**Tables 12–6 and 12–7**).

TABLE 12–6 Wheelchair (WC) Prescriptions for Spinal Cord Injury Patients Based on Motor Level

	CHAIR TYPE	ARMS	LEGS	PELVIC SEAT BELT/REAR WHEELS/TIRES	SEAT TYPE BOARD	CUSHION
High tetraplegia C2–C4	• Power WC with power tilt and/or recline • If ventilator dependent then ventilation tray • Control system depends on functional capabilities (i.e., sip and puff, chin control, head control) • High seat back, headrest, hip guides, and trunk supports	• Upper extremity support such as lap tray or arm trough with hand supports • Removable, flip-back, full-length armrests	• Leg rests: Elevating swing-away, removable, or center-mounted elevating; leg rests should be elevating if orthostasis is a concern • Swing-away footrests: With heel support loops—swing-away elevating leg rests should not be used for patients with significant hamstring tightness or environmental access issues	• Pelvic belt • Solid or pneumatic rubber tires, depending on patient's needs and caregiver maintenance capacity	• Solid seat	• Requires excellent pressure distributing seat cushion (i.e., 4-in. air-filled or viscous fluid with contoured foam
C5 tetraplegia	• Power WC with power tilt and/or recline • Modified joystick handle, midline mount joystick position may be necessary • High seat back, headrest, hip guides, and trunk supports	• Upper extremity support, such as lap tray or arm trough • Removable, flip-back, adjustable height armrests with either full-length or desk-length arm pads	• Leg rests: Elevating (to manage orthostasis), swing-away, removable, or center mount • Footrest with heel loop • See note above	• Pelvic (seat) belt • Solid or pneumatic rubber tires, depending on patient's needs and caregiver ability with maintenance • A manual WC with projections or coated handrim for backup to power WC or a power assist (see C6 manual following)	• Solid seat	• Requires excellent pressure distributing seat cushion (e.g., air-filled viscous fluid with contoured foam base) or 4-in. contoured foam cushion

(*continued*)

TABLE 12–6 Wheelchair (WC) Prescriptions for Spinal Cord Injury Patients Based on Motor Level (*continued*)

	CHAIR TYPE	ARMS	LEGS	PELVIC SEAT BELT/REAR WHEELS/TIRES	SEAT TYPE BOARD	CUSHION
C6 tetraplegia POWER (depending on strength and environment, can use either power or ultralight manual with power assist)	• Power WC • Joystick control with modified handle • High back seat or back support just below inferior angle of scapula—if power tilt or recline is not ordered	• Removable, flip-back desk-or full-length armrests	• Leg rests: Elevating swing-away, removable, or center mount elevating • Swing-away footrest with heel support loops • See notes above about hamstring consideration	• Pelvic (seat) belt • Solid or pneumatic rubber tires, depending on patient's needs and caregiver support	• Solid seat	• Requires excellent pressure relief seat cushion (i.e., air-filled viscous fluid with contoured foam base)
C6 tetraplegia MANUAL (can use either power or ultralight manual with power assist)	• Manual WC: Ultra-lightweight WC with solid back • Adjustable axle wheel positioning • Back height may be right below inferior angle of scapula • Brake extensions may be necessary	• Removable desk-length or tubular armrests	• Footrests: Swing-away with heel loops or rigid footrests with leg strap	• Pelvic belt • Solid rubber tires or high-pressure pneumatic tires with low rolling resistance • Modified handrims (i.e., projections or plastic coated) • Axle position can be placed more anterior on frame for improved maneuverability—Caution: Use anti-tippers until adequate WC skills are developed	• Solid seat	• Requires excellent pressure distributing seat cushion (i.e., air-filled, viscous fluid with contoured foam base 4-in. contoured foam cushion)
C7–T1 tetraplegia	• Manual WC: Ultra-lightweight with solid back • Adjustable axle wheel position • Back height may be right below inferior angle of scapula • Brake extensions initially	• Removable desk-length or tubular armrests	• Footrests—swing-away footrest with heel loops or rigid footplate with leg strap	• Pelvic (seat) belt • Solid rubber tires, Kevlar, or high-pressure pneumatic tires with low rolling resistance • Modified handrims (i.e., plastic coated) • Axle position can be placed more anterior on the frame for improved access and maneuverability—Caution: Use anti-tippers until WC skills are developed.	• Solid seat	• Requires excellent pressure distributing seat cushion (i.e., air-filled, viscous fluid with contoured foam base villous or 4-in. contoured foam)

TABLE 12–6 Wheelchair (WC) Prescriptions for Spinal Cord Injury Patients Based on Motor Level (*continued*)

	CHAIR TYPE	ARMS	LEGS	PELVIC SEAT BELT/REAR WHEELS/TIRES	SEAT TYPE BOARD	CUSHION
T2 paraplegia and below	• Manual WC: ultra-lightweight • Adjustable axle wheel position • Back height may be right below the inferior angle of the scapula or slightly lower due to better trunk stability	• Removable desk-length or tubular armrests; armrests should be chosen by patient preference; also, no armrests are an option	• Footrests—swing-away footrest with heel loops or rigid footplate with leg strap	• Pelvic (seat) belt • Solid rubber tires, Kevlar, or high-pressure pneumatic tires with lowest rolling resistance • Axle position can be placed more anterior on the frame for improved maneuverability—Caution: Anti-tippers necessary until WC skills are developed.	• Solid seat	• 📖 Requires very good to excellent pressure distribution seat cushion (i.e., 3- to 4-in. air-filled, viscous fluid with contoured foam or 3- to 4-in. contoured foam)

TABLE 12–7 Wheelchair (WC) Prescriptions for Hypothetical Patients

	CHAIR TYPE	ARMS	LEGS	SEAT BELT/TIRES	SEAT BOARD	CUSHION
Orthopedic patient with LE edema	• Standard WC—50 lb.	• Removable full-length or desk-length armrests	• Leg rests elevating, or footrest with heel loop • Swing-away	• Pelvic belt • Solid rubber tires	• Solid seat	• Contoured cushion with moderate pressure distributing cushion
Orthopedic patient S/P MI	• Lightweight WC—37 lb.	• Removable full-length or desk-length armrests	• Leg rests elevating or footrest with heel loop • Swing-away	• Pelvic (seat) belt • Solid rubber tires	• Solid seat	• Contoured cushion with moderate pressure distribution
Amputee	• Lightweight or ultralightweight WC with amputee axle • Wheels set back further (more posterior) to counterbalance loss of limb for more stable center of gravity	• Removable desk-length or full-length armrests if patient has difficulty transferring	• Amputated side: Amputee board • Unaffected side: Leg rests elevating; if necessary for circulation; otherwise, footrests with heel loops • Swing-away	• Pelvic (seat) belt • Solid rubber tires, pneumatic tires with airless inserts, or pneumatic tire	• Solid seat with amputee extension	• Contoured cushion with moderate pressure distribution quality cushion, if no significant skin risk

(*continued*)

TABLE 12–7 Wheelchair (WC) Prescriptions for Hypothetical Patients (*continued*)

	CHAIR TYPE	ARMS	LEGS	SEAT BELT/TIRES	SEAT BOARD	CUSHION
Hemiplegic patient	• Lightweight or ultra-lightweight WC with adjustable axle plate • Hemiplegic chair: This chair is lower, so the patient can self-propel with the uninvolved hand and foot (may need super low chair if patient is petite)	• Removable desk-length or full-length armrests if patient has difficulty transferring • Arm trough or half lap tray board (clear if left neglect present) • Brake extension on involved side	• *Hemiplegic side:* Leg rests elevating or footrest with heel loops • Swing-away • *Unaffected side:* None or swing-away footrest with heel loop when being propelled by others	• Pelvic belt • Pneumatic tires with airless inserts or pneumatic tires for traction and increased safety with transfers	• Solid seat	• Moderate to excellent pressure distributing cushion
General paraplegic athlete	• WC type will vary significantly for different sports. • Titanium, carbon fiber to ultralightweight WC—20–30 lb. • Wheel locks: Low mounted or scissor style (wheel locks set low for patients to obtain large push stroke) • Camber degree is selected according to sport • Anti-tippers are necessary in certain sports (front and/rear) • Racing WC may have three or four wheels	• Armrests are usually not utilized	• Footrests: Rigid footplate with leg strap	• Pelvic (seat) belt • Casters are smaller and narrower, making the chair more maneuverable • Solid rubber tires and higher air pressure pneumatic tires with low rolling resistance	• Solid seat	• Very good to excellent pressure distributing seat cushion

LE, lower extremity; S/P MI, status post myocardial infarction.

SINGLE SWITCH SCANNING

- Single switch control: The patient utilizes a single body movement such as eye blink or a puff of the cheek in order to activate a properly mounted single switch. The switch is typically paired with a scanner display module that is equipped with light-emitting diode lights or directional arrows that rotate through options, including forward/reverse, right/left, and mode select. Once the display module highlights the desired selection, the patient activates the switch to make a selection.
- Single switch driving is utilized with patients who have severe mobility or cognitive deficits after all other potential drive control methods have been ruled out.

BRAIN–COMPUTER INTERFACE CONTROL

- Brain–computer interface technology may serve as a potential method for wheelchair operation in individuals with severe neurologic disabilities.
- Possible input signals for brain–computer interface control include electroencephalography and event-related potentials (Kaufmann et al., 2014).

FUNDING

Many patients rely on an external funding source to help them obtain a wheelchair. Typical funding sources include private medical insurance, Medicare, Medicaid, Workers' Compensation, no-fault, and vocational rehabilitation programs. Depending on the funding source, there are specific documentation guidelines required to facilitate a patient's success with obtaining third-party funding for their wheelchair and seating system. Documentation often includes physician prescription, physician visit notes, and clinical documentation supporting the patient's need for a certain level of mobility product, seating system, and all the other essential components. Most medical documentation needs to be initiated and completed by the clinical team and other documentation needs to come from the supplier to the physician for review and signature.

CONCLUSION

This has been a basic overview of wheelchair prescription and measurement. It is important to remember that there are many additional components available to assist patients with ADLs, mobility, and everyday activities. These components are essential to help maximize a patient's ability to function as independently as possible at a wheelchair level. There are other seating systems available, such as scooters, which are not covered in this chapter. For further reading and a more in-depth review, please refer to the reading list at the end of this chapter.

OSTEOPOROSIS

Living bone is never metabolically at rest, as it is constantly remodeling and reappropriating its bony matrix and mineral stores along lines of mechanical stress. In the normal adult skeleton, the two processes of bone formation and bone resorption are coupled so that net bone formation equals net bone resorption **(Figure 12–10)**. Osteoporosis is a disease characterized by decreased bone mineral density (BMD; decreased bone mass/volume), which leads to decreased mechanical strength and an increased risk of fracture of the bone (Glaser & Kaplan, 1997). It is caused by an imbalance between bone formation and bone resorption.

WORLD HEALTH ORGANIZATION BONE MINERAL DENSITY CLASSIFICATION

The diagnosis of osteoporosis is made by measuring a patient's BMD and comparing it with the mean BMD of a young adult reference population with an average age of 30 years (World Health Organization [WHO] Scientific Group on the Prevention and Management of Osteoporosis, 2003).

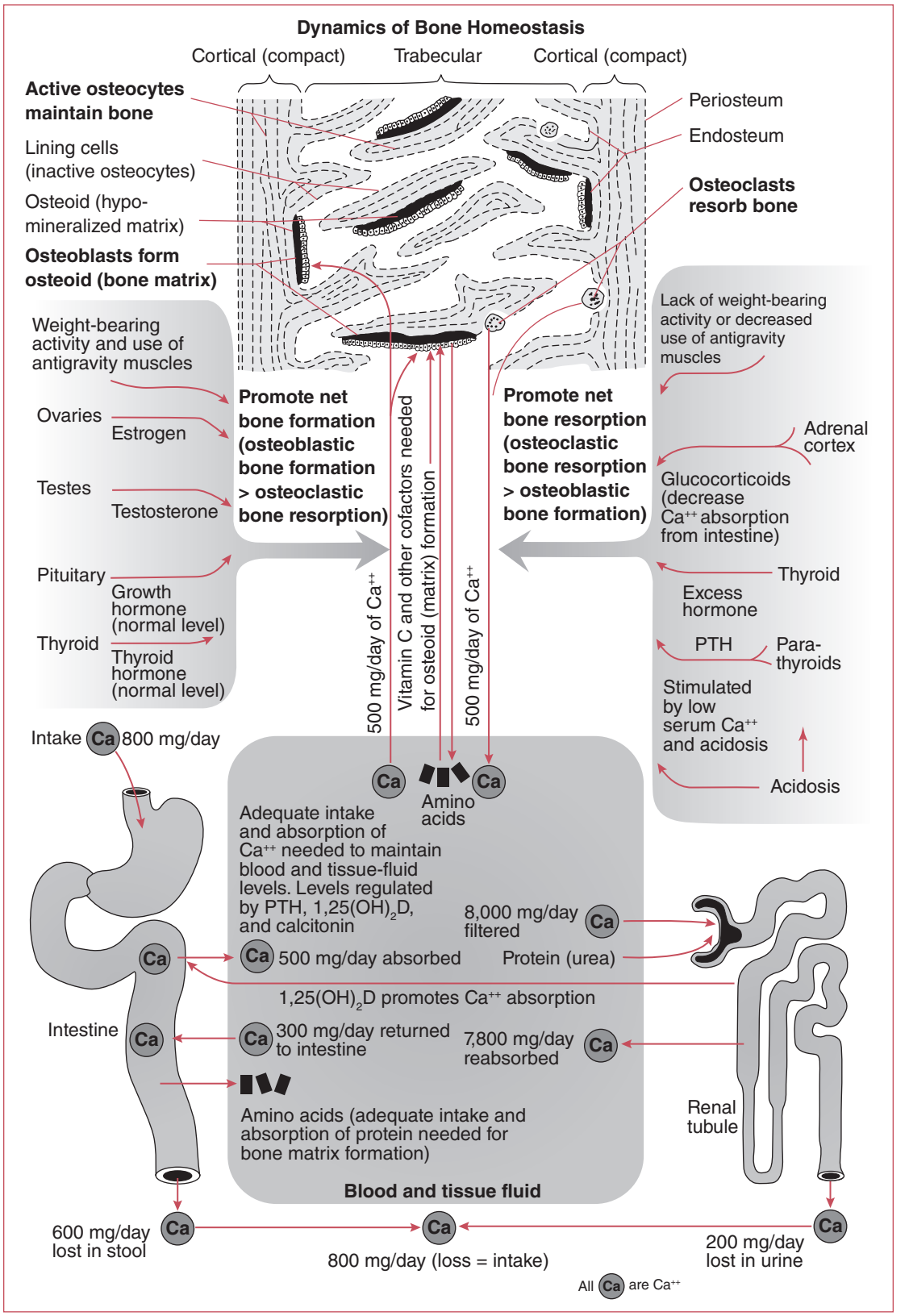


FIGURE 12–10 Calcium cycle.
PTH, parathyroid hormone.
Source: Redrawn from Kaplan FS. Prevention and management of osteoporosis. *Clin Symp*. 1995: 47(1):2–32.

One method called dual x-ray absorptiometry (DXA) scan (see Diagnosis of Osteoporosis section) is utilized to quantify BMD and is considered the gold standard for evaluating BMD. Results from a DXA scan are reported as *T*-scores and *Z*-scores. Diagnosis of osteopenia or osteoporosis is based on *T*-scores.

1. A ***T*-score** is the number of standard deviations (SDs) away from the mean peak bone mass (PBM) of young adults with the same race and gender (WHO Scientific Group on the Prevention and Management of Osteoporosis, 2003).
2. **Normal bone density:** Defined as a *T*-score between −1 SD and +1 SD (−1 SD ≤ normal *T*-score ≤ +1 SD).
3. **Osteopenia:** *T*-score is between −1 and −2.5, including −2.5.
4. **Osteoporosis:** *T*-score is <−2.5.
5. **Severe osteoporosis**: *T*-score is <−2.5, and the patient has suffered an osteoporosis-related fracture.
6. **Osteomalacia:** Defective skeletal mineralization of calcium or phosphate of bone osteoid (Fitzgerald, 2014).
7. A ***Z*-score** represents the number of SD the patient's bone density is in relation to adults of the same age, gender, and ethnicity (WHO Scientific Group on the Prevention and Management of Osteoporosis, 2003).
8. **Peak Bone Mass:** The highest level of bone mass achieved as a result of normal growth generally occurs between adolescence and age 35, with variation at specific skeletal sites.

T-SCORE:	–2.5		–1	0	1	2.5
	Osteoporosis	Osteopenia	Within normal range			

FACTS ABOUT OSTEOPOROSIS

- Most common metabolic bone disease
- Osteoporosis is a loss of osteoid bone that reduces structural bone integrity and results in an increased risk of fracture. It is usually asymptomatic until a fracture occurs (Fitzgerald, 2014). However, the diagnosis is not dependent on the patient suffering a fracture.
- Normal ratio of organic:mineral components but decreased density of bone tissue
  - Differs from osteomalacia, which has normal or increased bone tissue density but a reduced mineral content:organic component ratio
  - Osteoporosis and osteomalacia often coexist. Sun exposure and vitamin D supplementation are useful in preventing and treating osteomalacia but not osteoporosis (Fitzgerald, 2014).
- Major underlying cause of long bone fractures in the elderly is osteoporosis.

EPIDEMIOLOGY

- In the United States, 10.2 million adults have osteoporosis, and 43.3 million have bone mass levels that put them at risk for developing it (Sarafrazi et al., 2021). Of these, approximately 80% are women (International Osteoporosis Foundation, n.d.-a, n.d-b).
- Osteoporosis occurs more frequently with increasing age, as bone tissue is lost progressively (Lindsay & Cosman, 2014).
- Most women meet the diagnostic criterion for osteoporosis by age 70 to 80 due to loss of ovarian function at menopause precipitating rapid bone loss (Lindsay & Cosman, 2014).
- The prevalence of osteoporosis is increasing, most notably in women. From 2007 to 2008 through 2017 to 2018, the age-adjusted prevalence of osteoporosis in adults aged 50 and over increased from 9.4% to 12.6%, and from 14.0% to 19.6% in women (Sarafrazi et al., 2021). Prevalence is expected to continue to increase over successive decades (International Osteoporosis Foundation). There are about 1.5 million fractures per year in the United States related to osteoporosis, which accounts for a loss of millions of Disability-Adjusted Life Years

(DALYs) per year and imposed an economic burden of US$17 billion in 2005 (Burge, 2009; Fitzgerald, 2018; Johnell & Kanis 2006).

- In the United States and Europe, osteoporosis-related fractures are more common among women than men, with women accounting for about 75% of all hip fractures, which is likely due to lower PBM and postmenopausal bone loss (International Osteoporosis Foundation; Lindsay & Cosman, 2014).
- Worldwide, one in three women and one in five men over age 50 will experience osteoporotic fractures in their lifetime (International Osteoporosis Foundation).
- Approximately 70% of fractures in people >45 years old are related to osteoporosis.
- White women have a 40% lifetime risk of sustaining one or more fractures related to osteoporosis (Fitzgerald, 2014).
  - Hip and vertebral fracture incidence increases exponentially with age, with the peak age of hip fracture at 75 to 79 years old, while fractures of the distal radius dramatically increase in White women aged 45 to 60, then have a modest age-related increase (International Osteoporosis Foundation; Lindsay & Cosman, 2014).
- The risk of fracture is increased by 86% in people who have already sustained an osteoporotic fracture. 10% will have another fracture within 1 year, 18% within 2 years, and 31% within 3 years (International Osteoporosis Foundation).
- Hip fractures are a significant cause of morbidity and mortality in White women aged 50 years and above and, to a lesser extent, in White men of similar age.
  - The probability that a 50-year-old White individual will have a hip fracture during their lifetime is 14% for women and 5% for men, and incidence of hip fractures doubles every 5 years after age 70 (Lindsay & Cosman, 2014).
  - Fractures usually result in temporary disability, but approximately 50% of females with hip fractures are admitted to a nursing home; 14% of these patients remain in the nursing home after 1 year. Rates of nursing home admission rise with age (International Osteoporosis Foundation).
  - Morbidity: 50% of patients with hip fracture require assistance with ADLs; 15% to 25% receive long-term placement.
  - Mortality: 8% to 9% of patients die due to complications within 30 days and approximately 25% to 30% within 1 year (Luke & Ma, 2014). Women who have sustained a hip fracture have a 10% to 20% higher mortality than would be expected for their age.
- The National Osteoporosis Foundation uses the quality-adjusted life year of fractures to determine the impact of osteoporotic fractures on a person's life (Matkovic et al., 1996).

RISK FACTORS FOR OSTEOPOROSIS

Increased Risk

- Nonmodifiable factors:
  - Advanced age
  - Ethnicity: Caucasian or Asian
  - Female
  - Early menopause (<45 years) or bilateral oophorectomy
  - History of fracture as adult
  - Positive family history
  - Diminished PBM at skeletal maturity
  - Chronic inflammatory diseases (e.g., rheumatoid arthritis)
- Modifiable factors:
  - Malnutrition
  - Smoking
  - Excess alcohol intake
  - Estrogen/testosterone deficiency
  - Low calcium intake

- Inactivity/immobilization, impaired neuromuscular function
- Long-term medication use (e.g., high-dose glucocorticoids, proton pump inhibitors)
- Low body mass index (BMI), typically <20
- Female Triad—(see Female Athlete Triad Syndrome section later in this chapter)

Decreased Risk

- Obesity

PHYSIOLOGY

Cellular Components of Bone Remodeling

- **Osteoblasts:** Bone forming cells that produce an organic matrix, which is mineralized to form normal lamellar bone
- **Osteoclasts:** Bone resorption cells
- **Osteocytes:** Osteoblasts incorporated in new bone matrix

Skeletal Mass Components

CORTICAL BONE (COMPACT)

- Dense outer shell of bone
- Concentric lamellae, Haversian systems
- 80% of skeleton, most in shafts (diaphysis) of long bones
- Accounts for majority of skeletal mechanical strength

TRABECULAR BONE (CANCELLOUS, SPONGY)

- Found in central medullary canal; encloses marrow space
- Irregular branching plates, circumferential lamellae
- 20% of total skeleton, found in vertebrae and flat bones (axial skeleton), ends of long bones
- Vertebral bodies 42%, whole vertebrae 25%
- More metabolically active than cortical bone
- Preferentially altered in osteoporosis Type I

MINERALS, HORMONES, VITAMINS

- Calcium phosphate (in the form of hydroxyapatite):
  - Major component of bone
  - Regulated by parathyroid hormone (PTH), calcitonin, and vitamin D; 99% of body's calcium is in bone
- PTH regulated by Ca^+ concentration
  - Effects:
    - In bone: Activates osteoclasts
    - In intestine: Stimulates formation of vitamin D in kidneys (converts inactive form of vitamin D to active form of vitamin D), which results in increases in Ca^+ uptake
    - In kidneys: Increased phosphate excretion increases calcium reabsorption
- Calcitonin:
  - Synthesized by C-cells in thyroid glands
  - Decreases serum Ca^+ levels by inhibiting osteoclasts and increasing Ca^+ incorporation into bone
  - Calcitonin is U.S. FDA-approved for the treatment of osteoporosis in women >5 years postmenopause when alternative treatments are not suitable. It is also FDA-approved to treat Paget disease of bone and hypercalcemic emergencies (McLaughlin et al., 2024).
  - Not considered a first-line treatment for osteoporosis, as fracture reduction efficacy has not been demonstrated

  - The FDA notes that recent meta-analysis studies suggest an increased risk of malignancies with calcitonin-salmon use versus placebo, but no causal relationship. As a result, the need for continued use of calcitonin should be reevaluated periodically (Trovas et al., 2002; U.S. Food and Drug Administration, 2015).
- Vitamin D (1,25—Dihydroxyvitamin D_3):
  - In the intestines: Enhances Ca^+ and phosphorus absorption
  - In the kidneys: Increases reabsorption of Ca^+, phosphorus
  - 🕮 Vitamin D deficiency is the most common cause of osteomalacia.
  - Serum vitamin D, 25(OH)D3 levels <50 nmol/L or 20 ng/mL are generally considered inadequate for bone health (NIH Fact Sheet on Vitamin D).
  - Vitamin D deficiency correlates with musculoskeletal pain and is common in the inpatient rehabilitation setting (Heath, 2006; Nemunaitis et al., 2010; Neo & Kong, 2016).
    - Risk factors for vitamin D deficiency include advanced age, diminished exposure to sunlight, darker skin pigmentation, malnutrition, malabsorption, anticonvulsants, nephrotic syndrome, chronic kidney disease, and liver disease (Fitzgerald, 2014; NIH Fact Sheet on Vitamin D).
  - Most people in the United States consume less than the recommended amount of vitamin D.
  - In the American diet, vitamin D is found primarily in D3-fortified foods such as dairy products and in animal-based foods.
    - Animal products contain vitamin D3 and its three to five times more potent 25(OH)D3 form
      - Fatty fish and fish liver oils are among the richest sources.
    - Plant products such as mushrooms contain vitamin D2.
    - Both D2 and D3 supplements are well-absorbed in the gut, but D3 increases serum levels to a greater degree and for longer duration than D2.
  - In addition to dietary sources, sunlight exposure also contributes to vitamin D levels by catalyzing conversion of cutaneous D3 precursors to the active vitamin (NIH Fact Sheet on Vitamin D).

MAIN DETERMINANTS OF OSTEOPOROSIS

The pathogenesis of osteoporosis is multifactorial. Causes include genetic and environmental factors.

1. **Increased rate of bone loss:**
   - After PBM is achieved, bone loss occurs gradually thereafter, with the most rapid bone loss occurring in the early postmenopausal period in females.
   - Rate of age-related bone loss is approximately 0.25% to 1% per year in males and females.
   - Immediate postmenopausal period: 3% to 5% bone loss per year in females for approximately 5 to 7 years after onset of menopause.
   - Lifetime bone loss: 20% to 30% in males, 45% to 50% in females
   - Greatest contributor to bone loss in mature adults is loss of gonadal function.
2. **Quality of bone microstructures:**
   - Decreased trabecular bone (trabecular perforation)
3. **Failure to achieve adequate PBM at skeletal maturity:**
   - High-risk groups include persons such as the female athlete triad (see "Female Athlete Triad Syndrome" section later in this chapter), anorexics, persons with decreased mobility, and persons with nutritional deficiencies.

CLASSIFICATION

1. Generalized—Affects Different Parts of the Whole Skeleton

PRIMARY OSTEOPOROSIS (INVOLUTIONAL)—MOST COMMON

- Basic etiology is unknown.
- Postmenopausal osteoporosis (also known as Type I):
  - Typically during the 15 to 20 years following menopause

  - Affects females 50 to 65 years old—trabecular > cortical bone loss
  - Most fractures in spine, hip, and wrist (Colles fracture; i.e., axial skeleton)
- Senile or age-associated osteoporosis (also known as Type II):
  - >70 years old
  - 2:1 female:male ratio
  - Trabecular ≈ cortical bone loss
  - Fractures: Hip, spine, pelvis, humerus
- Juvenile: Children and adolescents; self-limited
- Idiopathic: Premenopausal females, middle-aged males

SECONDARY OSTEOPOROSIS (ALSO KNOWN AS TYPE III): ACQUIRED OR INHERITED DISEASES/MEDICATION-INDUCED

DISEASES

- Hyperparathyroidism
- Hyperthyroidism
- Cushing's disease
- Hypophosphatasia
- Hypogonadism
- Hypoestrogenism (anorexia, exercise-induced amenorrhea)
- Renal disease
- Chronic obstructive pulmonary disease (COPD)
- Systemic mastocytosis
- Rheumatoid arthritis
- Diabetes mellitus
- Idiopathic hypercalciuria
- Gastrointestinal (GI) disease (malabsorption syndromes, liver disease, partial gastrectomy)
- Alcoholism
- Nutrition (vitamin deficiency, calcium deficiency; high sodium, protein, phosphate, and caffeine intakes)
- Malignancy (multiple myeloma, lymphoma, leukemia)
- Immobility (tetraplegia/paraplegia/hemiplegia, prolonged bedrest)
- Loss of ovarian function → estrogen depletion, testosterone deficiency

MEDICATION-INDUCED

- Corticosteroids: Most common cause of secondary osteoporosis. Predominantly inhibits bone formation (↓ osteoblasts); mainly trabecular bone loss, which can lead to compression deformities of the vertebrae and pelvis.
- Heparin
- Anticonvulsants
- Excess tyrosine
- Lithium
- Loop diuretics

2. Localized—Discrete Regions of Reduced Bone Mass

PRIMARY

- Transient regional: Rare, migratory; predominantly involves hip; usually self-limited
- Complex regional pain syndrome Type I (formerly known as reflex sympathetic dystrophy): Radiographic changes may occur in the first 3 to 4 weeks, showing patchy demineralization of the affected area.

SECONDARY

- Immobilization, inflammation, tumors, necrosis

🕮 FEMALE ATHLETE TRIAD SYNDROME (Mcnamara & Walsh, 2014)

General

- Also known as the Female Triad, it consists of low energy availability (with or without an eating disorder), functional hypothalamic menstrual irregularity (amenorrhea or oligomenorrhea), and osteoporosis secondary to hypothalamic dysfunction → hypoestrogenism (Nazem & Ackerman, 2012).
- Higher rates seen in sports that emphasize leanness such as gymnastics, figure skating, dancers, distance runners, divers, and swimmers
- Disordered eating such as anorexia and bulimia may be seen.
- Skeletal demineralization can result in premature osteoporosis.

Clinical Features

- A history of stress fractures should raise concern.
- Diminished athletic performance may result.

Treatment

- Multidisciplinary approach is the most helpful, with the team including a physician, nutritionist, counselor, and coach or trainer.
- Calcium and vitamin D supplementation may be beneficial.
- Goals include improving caloric intake and possibly decreasing energy expenditure by limiting aerobic exercise.

DIAGNOSIS OF OSTEOPOROSIS

History and Physical Exam

- Evaluate for presence of risk factors or predisposing medical conditions.
- The first clinical indication is usually a fracture, typically the result of minimal trauma.
  - Fracture of proximal femur, distal forearm; pain usually present
  - Vertebral body fracture may be painful or asymptomatic; only one-third recognized clinically, and these are often incidental radiographic findings (Lindsay & Cosman, 2014).

Diagnostic Evaluation

- DXA scan
- Lab work: Complete blood count, comprehensive metabolic panel including liver function tests (LFTs), serum Ca^{+}, phosphorus, total alkaline phosphatase
- Fracture Risk Assessment Tool (FRAX®): A tool that evaluates an individual's 10-year probability of hip fracture and major osteoporotic fracture (in the spine, hip, shoulder, or forearm). It is valid only for postmenopausal women age 40 and older not currently taking prescription medications for osteoporosis (Menopause Society, 2024). It is available online.

Additional Diagnostic Studies

- Ionized Ca^{+}, elevated sedimentation rate, vitamin D, protein electrophoresis, thyroid function tests, PTH, follicle-stimulating hormone, estradiol, testosterone, serum and urine markers, urine Ca^{+}:creatinine ratio
- Iliac crest bone biopsy with tetracycline labeling (osteoporosis shows thin cortices and decreased number of trabeculae)
- **Markers of bone resorption:**
  - Elevated levels of calcium/creatinine ratio in fasting urine, hydroxyproline/creatinine ratio in fasting urine, collagen cross-links (pyridinolines, telopeptides), tartrate-resistant acid phosphatase

- **Markers for bone formation:**
  - Serum osteocalcin y-carboxyglutamic acid protein
  - Serum total and bone-specific alkaline phosphatase
  - Procollagen propeptide

Indications for Bone Mineral Density Measurement (National Osteoporosis Foundation, 1998)

- Women age 65 and older and men age 70 and older, regardless of clinical risk factors
- Younger, postmenopausal women and men age 50 to 70 who are of concern based on their clinical risk factor profile
- Women in the menopausal transition if there is a specific risk factor associated with increased fracture risk, such as low body weight, prior low-trauma fracture, or high-risk medication
- Adults who have a fracture after age 50
- Adults with a condition (e.g., rheumatoid arthritis) or taking a medication (e.g., glucocorticoids, 5 mg/day for 3 months) associated with low bone mass or bone loss
- Anyone being considered for pharmacologic therapy for osteoporosis
- Anyone being treated for osteoporosis to monitor treatment effect
- Anyone not receiving therapy in whom evidence of bone loss would lead to treatment
- Postmenopausal women discontinuing estrogen should be considered for bone density testing
- Estrogen-deficient women at clinical risk for osteoporosis
- Individuals with vertebral abnormalities
- Individuals receiving or planning to receive long-term glucocorticoid therapy 5 mg/day of prednisone or an equivalent dose for 3 months
- Individuals with primary hyperparathyroidism
- Individuals being monitored to assess the response or efficacy of an approved osteoporosis drug therapy

Other Indications

- If risk factors present for fractures in perimenopausal and postmenopausal women
- Screen for bone loss in conditions in which osteopenia is a manifestation
- Following response to treatment
- Testosterone-deficient men
- Research: Epidemiologic studies, clinical therapy trials

Radiographic Studies

🕮 DUAL X-RAY ABSORPTIOMETRY SCAN

- Gold standard for BMD measurement testing
- Accurate, precise, fast
- Low radiation exposure (scan times are shorter than with dual photon absorptiometry [DPA] and radiation dose is very low)
- Uses x-ray source instead of an isotope source
- There is suspicion because the radiation source does not decay and the energy stays constant over time.
- Allows assessment of axial or peripheral skeleton or entire skeleton
- Sites measured include the spine, hip, and radius.
- Proximal femur density measurement useful for predicting hip fractures
- Lumbar spine density measurement useful for monitoring response to therapy
- Spinal osteophytes and aortic calcifications may contribute to false high readings.

X-RAY

- Not sensitive in assessing bone mass
- 30% to 35% bone mass loss must occur before demineralization can be detected.

- *Findings:*
  - Cortical thinning
  - Trabecular pattern coarsened due to loss of small trabeculae
- *Findings in the spine:*
  - Increased radiolucency
  - Increased prominence of endplates
  - Increased concavity of endplates if nucleus pulposus has not degenerated (codfish vertebrae)
  - Anterior wedging and vertebral body height loss due to vertebral body compression fractures

SINGLE PHOTON ABSORPTIOMETRY/SINGLE X-RAY ABSORPTIOMETRY

- Inexpensive
- Low radiation dose
- Requires water bath or gel immersion
- Uses I^{125}, a radioactive isotope, in single photon absorptiometry (SPA) or x-ray source in single x-ray absorptiometry (SXA)
- Site measured: Radius, calcaneus
- Limited to bone measurement of peripheral skeleton; unable to measure bone density of hip or spine

DUAL PHOTON ABSORPTIOMETRY

- No water bath or immersion needed
- Uses AGD^{153} source, a radioactive isotope
- Less accurate and precise than DXA
- Increased scan time
- Sites measured: Proximal femur, lumbar spine

QUANTITATIVE COMPUTED TOMOGRAPHY

- Allows measurement of trabecular bone independent of spine, apart from cortical bone
- High-dose radiation and expensive
- Sites measured: Spine, hip, radius
- Accuracy compromised by increased fat content of bone marrow in elderly
- This technique is unique in that it provides for true 3D imaging and reports bone density measurement as true volume density measurements.
- The advantage of quantitative computed tomography (QCT) is its ability to isolate an area of interest from surrounding tissue.

QUANTITATIVE ULTRASONOGRAPHY

- Inexpensive, no radiation
- Not as precise as DXA. Diagnosis criteria for osteoporosis and treatment guidelines not well established with quantitative US.
- Sites measured: Calcaneus, tibia, patella, fingers

PHARMACOLOGIC TREATMENT

- Medications that preserve or improve bone mass
- Medications that decrease bone resorption

1. Calcium

- Calcium is important for optimal bone health. Although diet is the best way to get calcium, calcium supplements are an option if diet is deficient.
- Optimal calcium requirements are recommended by the National Institutes of Health (NIH) Consensus Panel **(Table 12–8).**

TABLE 12–8 NIH-Recommended Calcium Intake

AGE GROUP	RECOMMENDED CALCIUM INTAKE
Birth–6 months[a]	200 mg
7 months–1 year[a]	260 mg
1–3 years	700 mg
4–8 years	1,000 mg
9–13 years	1,300 mg
14–18 years	1,300 mg
19–50 years	1,000 mg
51–70 years	1,000 mg (Male) 1,200 mg (Female)
70+ years	1,200 mg
Pregnant and nursing women 14–18 years	1,300 mg
Pregnant and nursing women 19–50 years	1,000 mg

[a]Adequate intake.
NIH, National Institutes of Health.
Source: From National Institutes of Health, Office of Dietary Supplements. Calcium: factsheet for health professionals. Updated January 3, 2024. Accessed January 24, 2024. https://ods.od.nih.gov/factsheets/Calcium-HealthProfessional/.

- Immobilization and excess calcium intake predispose patients to kidney stones. Maintain urinary calcium excretion <250 mg per 24 hours in those without kidney stones.

2. Vitamin D

- Increases Ca^+ absorption in gut. Recommended doses: 400 to 800 International Units (IU) depending on age range (NIH Office of Dietary Supplements Fact Sheet, 2024).

3. Estrogen

- Mechanism of action:
  - Suppresses interleukin-6 secretion with inhibition of osteoclast recruitment
  - Decreases bone resorption
  - Increases calcium absorption in the gut
- Dosing regimens:
  - 0.625 to 1.25 mg/day conjugated estrogen cycled or continuous with progesterone 2.5 to 10 mg
  - Transdermal estradiol: 0.05 to 0.10 mg weekly
- Maintain therapy 10 to 20 years after onset of postmenopausal symptoms.
- Intact uterus: Use progesterone to decrease buildup of endometrium.
- If patient has had a hysterectomy, they may use estrogen only
- Benefits:
  - Preserves bone mass at multiple skeletal sites
  - Decreased incidence of fracture:
    - Approximately 50% risk reduction of spine fracture
    - Approximately 60% incidence reduction of hip/wrist fracture
  - Prevents vasomotor symptoms
- Side effects:
  - Endometrial cancer risk increased if estrogen is given without progesterone

  - Breast cancer if prolonged use after menopause
  - Increased risk of thromboembolic disease
  - Cardiac risks/benefits with use of estrogen remain controversial.
- Absolute contraindications: Breast cancer, estrogen-dependent neoplasia, thromboembolic disorder, hypercoagulable states, unexplained vaginal bleeding
- Relative contraindications: Uterine leiomyomas or cancer, familial hypertriglyceridemia, migraine, strong family history of breast cancer, endometriosis, chronic hepatic dysfunction, gallbladder disease

4. Calcitonin

- Directly inhibits osteoclastic activity, decreasing calcium levels in the serum. It also reduces calcium and phosphate levels in the blood through its effect on the renal tubules (McLaughlin et al., 2024). Derived from salmon.
- Benefits:
  - Decreases pain in acute compression fractures through stimulation of beta-endorphins
  - Preserves bone mass
  - Approximately 36% incidence reduction of spine fractures
  - As noted previously, more recent meta-analysis suggests increased risk of malignancies with calcitonin use versus placebo but no causal relationship.
- Recommended formulations:
  - Nasal spray (Miacalcin, Fortical®) 1 spray 200 IU/day, alternate nostrils
  - Parenteral injection subcutaneously or intramuscularly (Calcimar®): Prevention 100 IU every other day, treatment 100 IU daily for postmenopausal osteoporosis
  - Must have adequate concurrent intake of calcium and vitamin D
- Side effects: Nasal irritation, facial/hand flushing, local skin irritation, nausea; allergic reaction with injection form; hypersensitivity reaction (if using salmon calcitonin) including anaphylaxis, bronchospasm, swelling of the tongue, or rashes (McLaughlin et al., 2024)

5. Bisphosphonates

- Bisphosphonates have been shown to increase bone mass and reduce the incidence of spinal fractures. They must be taken in the morning with at least 8 oz of water 40 minutes prior to eating in an upright position to ensure proper absorption (Fitzgerald, 2014).
- Oral side effects: Abdominal pain, nausea, dyspepsia, difficulty swallowing, inflammation of the esophagus, and risk of ulceration, osteonecrosis of jaw, visual disturbances, and musculoskeletal pain
- IV side effects: Flu-like symptoms, fever, muscle and joint pain, headache, osteonecrosis of jaw, visual disturbances
- **Alendronate**(Fosamax®):
  - Recommended dosing: Prevention—5 mg/day; treatment—10 mg daily
    - Approximately 49% risk reduction of spine fractures
    - Approximately 56% risk reduction of hip fractures
- **Risedronate** (Actonel®):
  - Dosing prevention and treatment: 5 mg daily or 35 mg weekly or 75 mg twice monthly
  - Risk reduction of new spinal fractures up to 65%
- **Ibandronate** (Boniva®):
  - Dosing prevention and treatment: oral—2.5 mg daily or 150 mg monthly
  - 3 mg IV every 3 months
  - Comparable to Fosamax® and Actonel® in reducing vertebral fracture
- **Zoledronic acid** (Reclast®):
  - Dosing: 5 mg IV yearly; to be infused in no less than 15 minutes
  - Reduces vertebral fracture by 70%, hip fracture by 41%, and nonvertebral fracture by 25%

6. Teriparatide (Forteo®)

- Injectable form of recombinant human PTH
- Stimulates new bone formation in spine and hip
- For treatment of osteoporosis in postmenopausal women and men at high risk for fracture
- Recommended dosing: 20 µg subcutaneously daily; approved for use up to 24 months
- Side effects: Nausea, dizziness, leg cramps, cough

7. Selective Estrogen Receptor Modulators

- For prevention and treatment of osteoporosis in postmenopausal women unable to take estrogen due to side effects or risk of breast cancer
- 1% increase in BMD over 2 years in postmenopausal women compared with 2% with estrogen replacement therapy (Fitzgerald, 2014)
- **Raloxifene** (Evista®):
  - Dosing: 60 mg daily
  - Side effects: Hot flashes, increased risk of DVT

8. Increase Bone Formation (Positive Bone Formers)

- These medications are not FDA-approved for the treatment of osteoporosis.
- Sodium fluoride: Stimulates osteoblast formation; high dosage may increase risk of nonspinal fracture and bone fragility
- Anabolic steroids: May have beneficial effect on bone mass but side effects prohibit their use; side effects include nausea, GI bleeding, and joint pain
- Testosterone: May benefit men with hypogonadism

MANAGEMENT

Preventive Exercise

Activities involving weight bearing (axial loading) and resistance exercises (which allows the pull of functioning muscle) preserve or increase bone mass. They reduce the risk of osteoporosis by maximizing bone mass in young adults, maintaining and increasing bone mass in mature adults, and lessening bone loss in postmenopausal women.

ACTIVITIES ASSOCIATED WITH INCREASED BONE MASS (HIGH-IMPACT ACTIVITIES)

- Jogging or running
- Weight training
- Aerobics
- Stair climbing
- Racquet, field, and court sports
- Dancing

Therapeutic Exercise

The goals are to mitigate bone loss, increase strength and balance, prevent falls, and avoid fractures. Exercise regimens should be tailored to fitness levels and anticipated propensity to fracture or current fractures.

GOALS OF THERAPEUTIC EXERCISE

- Short term: Education of proper posture, body mechanics, increasing strength, and aerobic capacity
- Long term: Prevention of falls and fractures; proper nutrition, strength, aerobic capacity with adequate spine support, pain management, psychological support

EXERCISE PRINCIPLES

- Weight-bearing exercises improve bone density.
- Avoid spine flexion exercises in spinal osteoporosis, which may predispose to vertebral compression fracture.
- Posture correction: Avoid kyphotic posture.
- Pectoral stretching
- Strengthening: Back extension and isometric exercises to strengthen the lumbar region, abdomen, and upper and lower extremities
- Deep breathing exercises
- Balance and transfer training
- Proper lifting techniques and body mechanics

TYPES OF FRACTURES

Vertebral Body Compression Fractures

- Evaluation: History/physical exam, x-rays of spine, bone scan, CT scan
- Acute vertebral fractures may follow minor injury or physical activity.
- Most common osteoporotic fractures: Vertebrae > hip > wrist
- Vertebral compression fracture is the most common type of fracture seen in osteoporosis.
  - Most common site: Lower thoracic, upper lumbar areas
  - Typically involve the anterior part of the vertebral body, resulting in anterior wedging
  - Treatment: Restrict flexion-based activities and exercises, which load anterior vertebral body.
  - Consider spinal bracing.
- Retropulsed fragments:
  - Bony fragments from a vertebral body compression fracture can retropulse into the spinal canal. This can potentially result in spinal canal/spinal nerve compromise, which can cause back pain with neurologic symptoms.

Trabecular Microfractures

- Pain in the absence of gross fracture visible on x-ray. May be seen on CT.

Multilevel Vertebral Body Compression Fractures

- Multiple collapsed/anteriorly wedged vertebrae ⟶ kyphosis (Dowager's hump)
- Loss of height
- Abdominal protuberance, GI discomfort due to resulting thoracic kyphosis from multiple fractures
- Severe cases can lead to pulmonary insufficiency
- Can also lead to costal iliac impingement syndrome (see later)

FACET JOINT DISEASE

- Most prominent abnormality at vertebral collapse level with smaller lesions above and below level

Costal Iliac (Iliocostal) Impingement Syndrome

- Syndrome of lumbar pain due to mechanical irritation of the lower ribs against the iliac crest resulting from spinal deformity caused by multiple osteoporotic vertebral body compression fractures
- Increased pain with lateral rotation and bending
- Treatment:
  - Relief with soft wide belt, which sinks into the pelvic cavity, avoiding rib contact with the iliac crest
  - Injection of sclerosing material into the margins of the iliac and lower ribs
  - Resection of lower ribs

Limb Fractures

- **Hip fractures:** Multifactorial causes with two major risk factors: Osteoporosis and falls. Muscle forces acting on the hip are greater than the mechanical ability of the femur to withstand these forces.
  - Direction of fall is a major risk factor for fracture.
  - Elderly tend to fall sideways or drop in place.
- **Wrist fractures:** Most common fracture in females >75 years old

FALLS

Risk Factors for Falling

- Impaired vision (decreased vision, poor depth perception)
- Cognitive impairment
- Balance/gait abnormalities
- Weakness (i.e., inability to rise from a chair without using one's arms)
- LE disability/foot problems
- Peripheral neuropathy
- Myelomalacia
- Sedative use
- Polypharmacy
- Environmental (inadequate lighting, rugs, lack of railings)

Fall Prevention Program

- General conditioning exercises
- Improve balance
- Assistive devices: Canes, walkers, grab bars, tub benches
- Adequate shoe wear, avoid high heels
- Modification of meds
- Environmental modification: Adequate lighting, removal of throw rugs, handrails for stairs, ramps

TREATMENT OF VERTEBRAL BODY FRACTURES

Management of vertebral body fractures varies with acuity, location of fracture, and pain. Conservative approaches include bracing and PT.

Acute Vertebral Body Fracture

- Pain is usually severe, and most intense at fracture level
- Sharp pain increased with movement and alleviated with bedrest
- Severe pain lasting 2 to 3 weeks with decreased severity for 6 to 8 weeks
- May be asymptomatic

TREATMENT

- Relative rest
- Spinal orthosis: Soft corset for comfort. Rigid brace can be considered but often avoided to prevent disuse atrophy and discomfort from the brace itself.
- Physical modalities: Superficial heat or cold
- Analgesic use for comfort. Nonsteroidal anti-inflammatory drugs (NSAIDs) use controversial—may delay fracture healing.
- Spinal augmentation procedures—controversial (see next section) but can be considered in persistently symptomatic cases.
- Avoid constipation.
- Avoid exacerbating activity, in particular flexion-based movements.

Chronic Vertebral Body Fracture

- Pain will typically resolve after acute and subacute phases of fracture. Like acute compression fractures, they can be asymptomatic.
- Midthoracic region is the most common location.
- Can result in mechanical deformity or paraspinal muscle spasm
- Radiates laterally; associated with exertion
- Consider superimposed diagnosis, such as discogenic or facet-mediated pain.

TREATMENT

- Relative rest
- Analgesics: Nonnarcotic agents, calcitonin
- Soft spinal orthosis for comfort
- Assess ADLs, use of devices to avoid aggravation of pain.
- PT: Core muscle strengthening, posture, balance and flexibility, body mechanics
- Avoid flexion-based activities → increase vertebral compression forces.
- Physical modalities: Heat or cold, transcutaneous electrical nerve stimulation (TENS), acupuncture
- Behavioral modifications: Biofeedback, hypnosis, counseling

SPINAL AUGMENTATION PROCEDURES

Use of spinal augmentation procedures such as vertebroplasty or kyphoplasty for the treatment of acute osteoporotic vertebral compression fractures remains controversial. The current literature has not shown a significant benefit with such interventions. Furthermore, such procedures were associated with higher medical cost utilization than conservative therapy or placebo (Firanescu et al., 2018; Mathis et al., 2004; McCullough et al., 2013; Rousing et al., 2009).

Balloon Kyphoplasty

- A minimally invasive procedure with the goal of reestablishing vertebral body height and reducing pain
- **Technique:** Fluoroscopic guidance is used to place a hollow-bore needle into a vertebral body. A balloon tamp is inserted into a needle and inflated to restore bone height. Cement is then injected into the vertebral cavity, and the needle is then removed. The cement hardens in approximately 15 minutes. Pain usually improves within days.
- **Risks:**
  - Risks associated with local or general anesthesia
  - Infection—bleeding/hematoma
  - Cement leakage:
    - Extravasation into spinal canal ⟶ SCI or nerve root damage, paralysis
    - Pulmonary embolism
  - Venous air embolism
  - Spinal instability (Ohba et al., 2013): Studies have shown increased risk of adjacent level microfractures.

Vertebroplasty

A minimally invasive cementing procedure similar to kyphoplasty, but no balloon tamp is utilized to restore the vertebral body height. Instead, cement is injected directly into the vertebral body at higher pressures. Vertebroplasty is associated with similar risks as kyphoplasty.

SPINAL BRACING FOR VERTEBRAL BODY FRACTURES

Indications for Spinal Bracing

- Pain relief: For acute fractures, spinal immobilization decreases paraspinal muscle spasm and overuse.
- Spinal stabilization

- Prevent further fracture
- Prevent soft tissue shortening
- Decrease flexion
- Compensate for weak erector spinal muscles

Contraindications

- Hiatal hernia
- Inguinal hernia
- Orthopnea secondary to COPD
- Obesity
- Kyphoscoliosis

Risks of Prolonged Use of Orthosis

- Weakening/atrophy of trunk muscles
- Reduced spinal mobility
- Increased fracture risk due to disuse osteoporosis

Types of Orthoses (see also Chapter 6, Prosthetics and Orthotics)

1. NONRIGID BRACE

- Abdominal corset (elastic binder): Helps to decrease pain. Increases intra-abdominal pressure and reduces axial load on vertebral bodies; also serves as a reminder to restrict motion.
- Used in stable fractures for pain management and in patients who are unable to tolerate rigid bracing (but do not have an unstable fracture)

2. RIGID BRACING

- Can be used in acute thoracic and lumbar (TL) vertebral body fractures but causes increased discomfort and noncompliance compared with soft bracing
- Thoracolumbar brace: Assist spine extension and prevent spinal flexion movement via shoulder straps and paraspinal bars; increases intra-abdominal pressure
- Thoracolumbosacral orthosis (TLSO):
  - Fixation from pelvis to shoulders
  - Most restrictive/greatest immobility
- Jewett brace: Forces act to extend thoracolumbar region
- ***C***ruciform ***A***nterior ***S***ternal ***H***yperextension (CASH) brace

Note: Orthotics that cause excessive hyperextension forces on the spine may induce posterior element type of pain or fractures in the osteoporotic patient. This therefore should be a consideration in this patient population.

Other

- **Postural training supports** consist of a brace suspended by loops from the shoulders with small pouches containing weights up to 2 lb. The weights are positioned just below the inferior angle of the scapula to counteract the tendency to bend forward and may be worn for 1 hour twice daily.

REHABILITATION OF BURN INJURIES

BACKGROUND

General

- A burn is the body's response to a soft tissue insult from an external agent, such as heat, cold, chemicals, electricity, or radiation.
  - 85% to 95% of burns are caused by heat.

  - 5% to 15% of burns are from frostbite, chemical, and electrical damage.
- 1.5 to 2 million people sustain burns each year in the United States.
- 450,000 burn victims receive some form of medical treatment, and approximately 30,000 burn victims need hospitalization in the United States.
- 35,000 to 50,000 people annually have temporary or permanent disability secondary to burns.
- Deaths caused by burn injury are the fifth most common cause of unintentional injury deaths in the United States (Esselman, 2007).
  - 5,000 people die each year from burns. Survival rate is >95%.
  - Mortality has declined because of the development of comprehensive burn centers.
- 🕮 Burns are:
  - #1 cause of accidental deaths in children under the age of 2. The majority of burns in this age group occur as a result of abuse.
  - #2 cause of accidental deaths in children under 4.
  - #3 cause of accidental deaths in children under 19.

Local Physiologic Response to Burns

- Local reactions to burns include:
  - Intense vasoconstriction secondary to epinephrine, prostaglandins, serotonin, and leukotrienes
  - Early vasodilation and increased capillary permeability secondary to histamine release. Within a few hours, histamine release causes vasodilatation and increased capillary permeability, allowing protein and albumin into the extravascular space. This is followed by fluid extravasation (→ fluid shifts), which can cause severe edema and hypovolemia.
  - Late capillary permeability secondary to leukotrienes
  - Swelling and rupture of damaged cells
- Fluid shifts can occur due to increased microvascular permeability and the development of a negative interstitial fluid pressure (secondary to a disruption in the capillary endothelial integrity). Fluid shifts from plasma into the interstitial space can result in edema and, in more severe cases, hypovolemia.
  - Exposed collagen causes platelet activation. Platelet and leukocyte aggregation with clot formation occurs from tissue thromboplastin, endotoxin, interleukin-1, and Hageman factor.
  - In severe burns, inflammatory mediator release occurs, which can lead to systemic symptoms such as shock.
  - Establishment of a hypermetabolic state develops.
- 🕮 Inflammatory mediators released upon thermal injury:
  - Histamine—causes an increase in arteriolar dilatation and tissue pressure, leading to increased microvascular permeability.
  - Prostaglandins—released from macrophages and neutrophils. Act as vasodilators and increase microvascular permeability.
  - Thromboxane—produced by platelets. Minimal effect on vascular permeability but can increase ischemia to the wound.
  - Catecholamines (epinephrine and norepinephrine)—cause arteriolar vasoconstriction, which is beneficial to reduce edema. However, the effects are not seen in injured tissues and are also offset by the opposite effects of the other inflammatory mediators.
    - Other vasoconstrictors: Angiotensin II, vasopressin, and platelet aggregation factor.

Systemic Physiologic Response to Burns

- Loss of fluid into the extravascular compartment results in hypovolemia and shock. Increased response from severe burns results in release of inflammatory mediators, which can also lead to shock (Schneider & Spires, 2010).

- Hyperventilation with increased oxygen demand
- Inhalational injury causing decreased oxygenation and acute respiratory distress syndrome (ARDS)
- Initial decrease followed by a significant increase in cardiac output after several days
- Increase in blood viscosity
- Gastric dilation and ileus occur in the first 3 days postburn.
- Multiorgan system failure in severe cases

CLASSIFICATION OF BURNS

Causative Agent

1. THERMAL BURNS

- Heat: Creates a zone of coagulation that destroys tissue and creates a zone of stasis, an area of decreased blood flow. This area may improve or get worse depending on treatment.
- Cold: Damage occurs as a combination of actual freezing plus decreased blood flow and ischemia. Alcohol is commonly involved in these injuries.

2. ELECTRICAL BURNS

- Superficial damage may appear minimal, but deeper tissues (muscle and bone) may incur severe injuries.
- Low-voltage currents travel through the body following the path of least resistance (i.e., nerves, arteries, veins, soft tissue) and can cause damage all along its course. High-voltage currents take a more direct path between entrance and ground.
- Currents are concentrated at the areas of greatest resistance (i.e., entrance and exit points to the body) with a maximal release in energy in those areas. As a result, injuries tend to be worse at those points. Exit wounds are often worse than entrance wounds.
- Injuries observed in conjunction with electrical burns include:
  - Radiculopathy caused by tonic–clonic contractions during electrocution
  - Peripheral neuropathy caused by direct injury from the current
  - Cognitive impairment
  - SCI
  - Formation of heterotopic bone around joints and in residual limbs; typically has a late onset of presentation
  - Cardiopulmonary arrest
  - Will be at risk of developing early onset of cataracts and hearing loss, both amenable to usual treatments

3. CHEMICAL BURNS

- From either acid or alkali exposure
- These burns are typically underestimated and will frequently appear to be mild in severity. Inappropriate or insufficient removal of the causative agent allows the injury to progress.

4. RADIATION BURNS

- Risk and severity of burn will depend upon the duration and intensity of exposure.
  - Response will vary from mild erythema to blistering and skin sloughing over a period of hours to days.
  - If exposure is high enough, treatment can only be palliative.
- The most common cause is sunburn. The next most common is radiation therapy; 95% of cancer patients receiving radiation therapy will develop some form of radiodermatitis.

Burn Injury Classification (Table 12–9)

Layers of the Skin

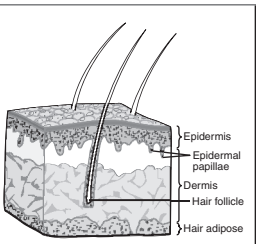
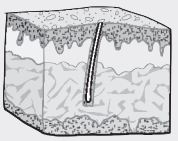
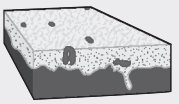
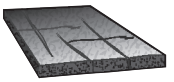
Epidermis	• Thin, outermost layer consisting of five layers: – Stratum corneum – Stratum lucidum – Stratum granulosum – Stratum spinosum – Stratum basale
Dermis	• Papillary layer (superficial) • Reticular layer (deep)
SC tissue	• Consists primarily of loose connective tissue and fat

SC, subcutaneous.

TRADITIONAL BURN CLASSIFICATION

- **First-degree burn:** Injury to outer layer of the epidermis, no injury to the dermis; erythema but no blistering
- **Second-degree burn:** Injury to full epidermis + superficial layer of the dermis, but deep layer of the dermis remains; blistering.
- **Third-degree burn:** Injury to full epidermis + most of dermis; white eschar due to compromised blood flow
- **Fourth-degree burn:** Destruction of all skin layers + subdermal tissue injury to fat/connective tissue, muscle, nerve, and bone

TABLE 12–9 Burn Classifications

NORMAL SKIN	Epidermis and dermis intact.	
SUPERFICIAL (First Degree)	Only the epidermis is injured, sparing deeper skin layers. Erythematous but no blistering.	
PARTIAL THICKNESS: SUPERFICIAL AND DEEP PARTIAL THICKNESS (Second Degree)	Involves epidermis + dermis; blistering is present. Superficial: Epidermis and upper dermis Deep: Extend into deeper dermis (haia follicles and glandular tissue)	
FULL THICKNESS (Third Degree)	Involves the full layer of epidermis and part of the dermis; extends through entire dermis; only the basal layer lining skin appendages remains.	
FULL THICKNESS + UNDERLYING TISSUE (Fourth Degree)	Total destruction of epidermis and dermis. Extends through the entire skin, into underlying fat, muscle and bone.	

Note: The degree of burn describes the depth of injury. Most injuries are of varying depths.
Source: From O'Young B, Young MA, Stiens SA. *Physical Medicine and Rehabilitation Secrets*. Hanley and Belfus; 1997, with permission.

NEW CLASSIFICATION

- **Superficial Burn:** Involves the epidermis only.
- **Partial Thickness Burns:**
  - **Superficial partial thickness:** Epidermis and upper third of dermis injured
  - **Deep partial thickness:** Epidermis and most of dermis injured
- **Full Thickness:** All layers destroyed

Size of Burn: Rule of 9s (Figure 12–11)

- The Rule of 9s is an *approximate* way of estimating adult total body surface area (BSA).
  - Head = 9% BSA
  - Each upper extremity = 9% BSA
  - Each LE = 18% BSA
  - Anterior trunk = 18% BSA
  - Posterior trunk = 18% BSA
  - Perineum = 1% BSA

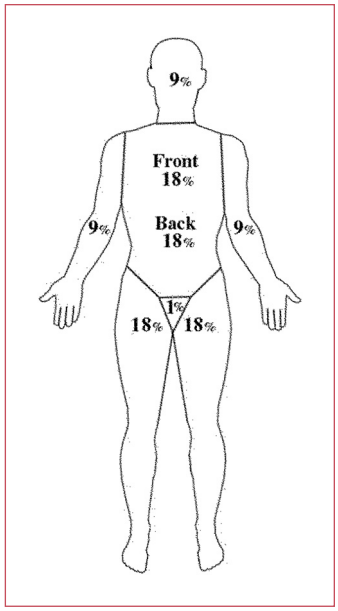


FIGURE 12–11 Rule of 9s in adults.

American Burn Association Classification of Burn Severity

1. **Minor:**
   a. <15% BSA partial thickness (10% in child)
   b. <2% BSA full thickness (not involving eyes, ears, face, or perineum)
2. **Moderate (most should be hospitalized):**
   a. 15% to 20% BSA (10%–20% in child)
   b. 2% to 10% BSA full thickness (not involving eyes, ears, face, or perineum)
3. **Major (all should be hospitalized):**
   a. >25% BSA partial thickness (20% in child)
   b. ≥10% BSA full thickness
   c. All burns to eyes, ears, face, or perineum
   d. All electrical burns
   e. All inhalation burns
   f. All burns with fracture or major tissue trauma
   g. All with poor risk secondary to age or illness

FACTORS AFFECTING OUTCOME

1. **Age:** Infants, children, and the elderly have a worse rate of survival.
2. **Total BSA:** The greater the area of involvement, the worse the prognosis. Of note, children have a different distribution of BSA because in an infant, the head is a much larger portion of the whole body compared with an adult. An adult's head comprises approximately 9% (rules of 9) of the total BSA, whereas a newborn's head comprises 19% of its total BSA. Thighs and legs will comprise 11% and 10%, respectively, of a newborn's total BSA and will increase with age to comprise approximately 18% and approximately 18% (rules of 9), respectively, in adults.
3. **Depth of burn:** As the total BSA that is full thickness increases, the prognosis decreases.
4. **Other associated injuries:** Prognosis decreases as the number of concurrent injuries increases.

MEDICAL MANAGEMENT OF BURN INJURIES

Initial Treatment

Always begin any emergency assessment with an evaluation of the ABCs (Airway, Breathing, Circulation). Special considerations include:

- Fluid resuscitation—use Brooke, Evans, Baxter, or Parkland formula as a guide

- **Parkland formula:** 4 mL/kg body weight divided by % BSA burned = half of the total calculated fluid requirement should be given in the first 8 hours. The remaining amount is divided equally over the next 16 hours. Most important physiologic parameter to follow is urine output: 0.5 to 1.0 mL/kg/hr.
- If fluid resuscitation is too aggressive, there can be a risk of developing compartment syndrome (Gabriel & Holavanahalli, 2016).
- Escharotomy: An incision of burned tissue to relieve pressure from edema and avoid neurovascular compromise and amputation
- Limbs requiring escharotomies should be elevated and splinted in neutral position for 24 hours (Gabriel & Holavanahalli, 2016).
- Nasogastric tube for abdominal decompression and Foley catheter for volume management
- Tetanus toxoid vaccination
- Cleaning all wounds with sterile saline
- Application of topical antimicrobials. Systemic antibiotics should await specific indications and be culture-driven. Systemic antibiotics are not recommended for prophylaxis of infection in majority of burn patients (Ramos et al., 2017).
- Rapid and extensive debridement and grafting. The goal is to cover as much open area as possible in as short a period of time as possible. Nothing else will prevent complications, decrease pain, and promote rapid recovery as well as coverage of wounds.
- Once the patient is stabilized, early mobility and exercise are important to initiate in order to maintain function (Gabriel & Holavanahalli, 2016). Physiatry consultation during acute care has been shown to decrease length of stay.

Wound Healing

PHASES

1. Inflammatory phase: Response to injury with influx of neutrophils and macrophages.
2. Proliferative phase: New matrix is laid with fibroblasts and in-growth of capillaries.
3. Maturation phase: Resolution of inflammation in which collagen is laid down to form the scar.
4. Epithelialization: Reestablishment of the basement membrane and epidermis.
5. Wound contraction: The open margins are brought together by fibroblasts.
6. Wound contracture: Shrinkage of the scar through collagen remodeling. This causes functional restriction of motion.

TREATMENT (TABLE 12–10)

- Wounds are more likely to have hypertrophic scarring if they take longer than 2 weeks to heal. Hypertrophic scarring will appear as red, raised, and rigid tissue that will contract and limit motion of the skin. To reduce this, early debridement and skin grafting with good local wound care is required (Esselman, 2007).
- **Wound dressings:** First use topical antimicrobial agents (e.g., Silvadene® or Bactroban®), then biological or synthetic dressings.
- **Biological dressings:** Temporary cover for a burn wound to protect the wound and decrease fluid loss from the site, decrease pain, and inhibit bacterial growth. May use autografts—split thickness skin grafts (STSG) or full thickness skin grafts (FTSG) taken from donor sites of unburned skin on the patient; homographs—skin from the same species but not the patient (human cadavers or fetal membranes); and xenografts—skin from other species (pigs and tilapia in the United States or frogs in Brazil). Homographs and xenografts are temporary coverings because the body will ultimately reject the foreign protein. They are utilized when too much BSA is involved and there is not enough uninjured skin to use for grafts, or until donor sites can be reharvested. Until then, they provide the same benefits as STSG or FTSG.
- **Synthetic dressings:** Significant effort has gone into the development of synthetic membranes. These are skin substitutes that can be used to engraft a wound site, ideally replacing the need

for an autograft, or can act as a temporary covering until an autograft is applicable. They may augment wound healing with biologic components including fibroblasts and keratinocytes. There are several products commercially available.

- **Cultured epithelial autocytes (CEA):** CEA is skin that is cloned from a small, 2.5-cm square piece of the patient's own skin. From that, literally yards of skin can be grown rapidly. It is the patient's own skin, so risk of rejection is minimized. Problems include fragility (making application technically very difficult), lack of a basement membrane (it is only the epidermal layer, meaning it cannot cover irregular surfaces well and easily slides off even after several days), and horrendous expense.
- **Debridement:** Removal of eschar to expose viable tissue and prepare the wound bed for coverage
  - **Mechanical:** Wet to moist dressings, or hydrotherapy to soften eschar
  - **Hydrotherapy:** Daily cleansing of wounds. Immersion tanks are not used secondary to the risk of cross-contamination.
  - **Enzymatic:** Digest necrotic tissue without harming viable tissue. May be painful, increase body temperature, or cause bleeding. It can increase fluid loss; therefore, it should only be done for 20% or less of BSA.
- **Surgical debridement:** Tangential excision: 1 to 10 days postburn, removing thin layers until normal viable tissue is exposed. May have significant bleeding. With early debridement and grafting, it has been shown to decrease hospital stay, mortality, and sepsis (Helm et al., 1998).
- **Skin grafting:** Covering a wound with healthy skin
  - Used if wound is not expected to heal within 18 to 21 days
  - Allows early wound closure, which reduces pain
  - Autologous
  - FTSG utilizes all layers of epidermis and dermis. It will not contract as it matures. It can only be used to cover small areas that are burned, as available tissue is rapidly used up.
  - STSG is meshed at the time of harvest, giving the largest possible area of coverage. Will contract as it heals.
  - Homologous (cadaver) split skin graft (temporary)
  - Xenograft donor graft from another species (temporary)
  - Immobilize the joint above and below the graft area for a minimum of 3 to 5 days to encourage healing.
  - Minimize positional-dependent edema to prevent graft loss.
    - Unless LE grafting crosses a joint, early mobilization is appropriate. Studies have shown no difference in graft loss between patients mobilized as early as day 1 postop compared with day 5.

REHABILITATION ISSUES

Contractures

- Prevalence of contractures is 40% to 55% at discharge from acute care for burn survivors (Dodd et al., 2017).
- Scars will grow and then contract. They will continue to contract unless they are met by an opposing force. Preventing and treating burn scar contractions require low-load, prolonged active and passive stretching and positioning. Scars usually mature in 1 to 1.5 years, but are occasionally active for much longer periods.
- Hypertrophic scars result from deep partial and full thickness burns (Schneider & Spires, 2010).
- Prevalence is 32% to 67% with severe burn injuries (Esselman, 2007).
- Hypertrophic scarring is associated with exposure to the fat domes in the deep dermis. The development of hypertrophic scarring is postulated to be due to altered interaction between adipose-derived stem cells and deep burn exudate (Chiang et al., 2016).
- Keloids and hypertrophic scars are similar. However, keloids will extend beyond the original injury and invade into local soft tissues (Gabriel & Holavanahalli, 2016).

TABLE 12–10 Assessment and Treatment of Burn Injuries

DEPTH OF INJURY	HEALING TIME	PAIN	WOUND OUTCOME	TREATMENT MODALITIES
Superficial epidermis (first degree)	• 1–5 days	• Painful for 1–3 days, ibuprofen or acetaminophen gives adequate analgesia	• No sequelae	• Elevation decreases pain of limb • Keep wound clean • Aloe or other moisturizer reduces dry skin and itching • If needed (usually in electrical injuries), therapy to prevent PTSD
Superficial dermis (second degree/superficial partial thickness)	• 14 days	• Painful for 5–14 days • Acetaminophen with codeine or oxycodone gives adequate analgesia for wound care, exercise, and sleep	• Possible pigment changes	• Wound care • Active exercise • Protective garments • Sunscreen • Therapy to prevent PTSD
Deep reticular dermis (second degree/ deep partial thickness)	• 21 days for spontaneous healing • If grafted after 10–14 days, less scar formation will be noted, with improved functional outcome; less pain and shortened hospital stay	• Very painful until closure • Methadone or oral morphine continuously for baseline pain control • Parenteral or instant-release oral morphine and/or oxazepam and midazolam for dressing changes and stretching exercises	• Probable pigment changes • Reduced skin durability • Severe scarring • Sensory changes • Apocrine changes • Edema in dependent limbs	• Wound care • Anti-inflammatories, analgesics • Antipruritics • Active exercise • Elevated positioning/orthotics • External vascular support garments • Moisturization and lubrication • Daily living skills • Psychological therapy • Therapy to prevent PTSD
Subcutaneous tissue (third degree/full thickness)	• Graft needed, or, if smaller, undermine to approximate with primary closure • Variable healing time	• Nonpainful initially due to destruction of nerve endings • Pain medication as above mentioned • Carbamazepine, phenytoin, or amitriptyline	• Same as above-mentioned • Additional sweating loss • Possible loss of finger or toenails • Possible additional sensory loss • Alopecia over grafts • Areas of cultured epithelial autograft show permanent fragility, loss of temperature control, dry blistering skin with changed sensation	• Same as above-mentioned • Postop positioning/immobilization • Possible need for NSAIDs or other etidronate disodium to prevent HO (controversial early treatment) • Therapy to prevent PTSD • Very slow weaning from analgesics and anxiolytics • Vibration for pruritus
Muscle, tendon, bone (fourth degree; old term in disfavor and rarely used)	• Amputation or reconstructive surgery, such as flaps, needed • Healing time variable	• Nonpainful initially due to destruction of nerve endings • Chronic pain treatment for neuromas and phantom limb pain and later bone spicules	• Variable outcome • Early amputation with closure using noninjured tissue shortens hospital stay, decreases pain, and improves prosthesis fit	• Same as above-mentioned • Deep tendon massage • Adapted equipment • Prosthetic fitting if indicated

Note: Ibuprofen has the dual action of inflammation reduction at injury site and pain reception reduced at the CNS level.
CNS, central nervous system; HO, heterotopic ossification; NSAIDs, nonsteroidal anti-inflammatory drugs; PTSD, posttraumatic stress disorder.
Source: Adapted from Rivers EA, Fisher SV. Burn rehabilitation. In: O'Young B, Young MA, Stiens SA, eds. *PM&R Secrets.* Hanley & Belfus; 1997419–420. with permission.

TABLE 12–11 Consequences of Burn Injury by Depth of Burn

SKIN LAYER	ABSENT OR IMPAIRED MORPHOLOGY	WOUND CONSEQUENCES
Epidermis	Stratum basale Stratum spinosum Stratum granulosum Stratum lucidum Stratum corneum Melanocytes	Source for proliferating cells Decreased protection Increased water loss Water loss, microorganism growth Entry of noxious agents Repeated sunburn
Dermis (does not regenerate)	Altered collagen Increased collagen Aging collagen	Decreased tensile strength Increased scarring (hypertrophic scarring) Altered surgical response
Nerves	Affected Absent	Pruritus/paresthesias Decreased sensation, trauma, and burn risk
Vascular system	Impaired Absent fragility	Impaired (especially venous return) No healing (depends on area) Reinjury risk
Basement membrane zone	Basal decidua and densa Rete pegs and dermal papillae	Blisters Blisters, fragility
Epidermal appendages	Sweat ducts Sebaceous glands Hair follicle	Impaired thermoregulation Loss of duct, sweat, and oil glands Loss of hair root, resultant alopecia
Fingernail bed	Basal cells for proliferation absent	Malformed or absent nail

Source: Adapted from Campbell MK, Covey MH, eds. *Topics in Acute Care and Trauma Rehabilitation.* Aspen Publishers; 1987, with permission.

- Contractures are particularly damaging over joints. Hypertrophic scarring is cosmetically and psychologically devastating even in the absence of mechanical limitation. 25 mmHg of pressure is needed to counteract the contraction of a scar.

POSITIONING TO PREVENT CONTRACTURE (FIGURE 12–12)

- Position the patient in extension and abduction. Patients tend to contract in flexion and adduction for comfort because those positions reduce stretching of the injured skin.
- Position to prevent dependent edema
- Use of special beds (e.g., Kin Air® and Roho) to limit pressure, breakdown, and facilitate positioning
- Positioning to avoid to minimize/prevent neuropathy:
  - Brachial plexus—avoid shoulder abduction >90 degrees and external rotation. The shoulder can be flexed in 90 degrees with 10 degrees of horizontal adduction to take off stress from brachial plexus.
  - Ulnar nerve—avoid elbow flexion at 90 degrees, upper extremity pronation.
  - Median nerve—avoid wrist hyperextension.
  - Peroneal nerve—avoid frog leg position, in which hip is abducted at 45 degrees and knee flexed approximately 30 to 40 degrees (Gabriel & Holavanahalli, 2016; Schneider & Spires, 2010).

SPLINTING

- Orthotic use after burns reduces contractures and improves ROM in adult burn survivors (Parry et al., 2020).

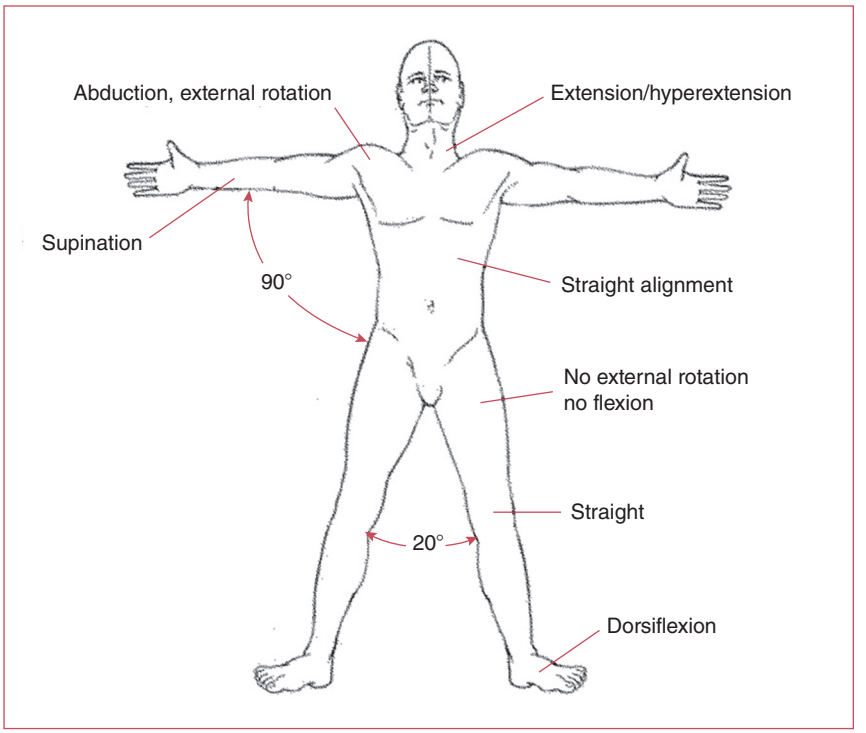


FIGURE 12–12 Anti-contracture positioning—patient supine from ventral position.

- Used with patients who are not compliant with positioning or if exposed tendons or joints are present
- Static, dynamic, and static progressive splints may be used. There is no definitive best-practice guideline for type of splint used.
- Splinting should not be performed without mobilization. An elbow that is frozen in full extension because it was splinted and never ranged is just as impaired as one that has contracted into full flexion because it was never splinted initially.
- 🕮 Splint positioning for elbow burns:
  - Elbow extended and forearm supinated
- 🕮 Splint positioning for dorsal hand burns:
  - Wrist in 15 to 20 degrees of extension
  - Metacarpophalangeal (MCP) in 60 to 70 degrees of MCP flexion
  - Proximal interphalangeal and distal interphalangeal placed in full extension—Thumb—slight interphalangeal (IP) flexion and palmar abduction
- 🕮 Splinting should be prescribed for exposed joints and tendons:
  - Splinting exposed joints is important to protect from ankylosis of the joint capsule.
  - Exposed tendons are splinted in the slack position to allow revascularization (see resting hand splint given later).
  - Resting hand splints are used for proper positioning in patients with general hand burns.
    - For hand burns with extensor tendons exposed, put the tendons on slack to prevent central slip failure that can result in a Boutonnière deformity.
- Splinting of LE burns:
  - Knee extension with foot dorsiflexion

TRANSPARENT FACE MASK—MADE OF SILICONE AND USED OVER HEALED GRAFTS

- Monitor skin more easily than fabric mask
- Custom-molded from the patient's own face for individual fit
- Total contact face mask orthoses protect and preserve the contours of the face.

- Prevents severe cosmetic deformities with even pressure across uneven surfaces (e.g., the nose)
- Clear mask is more cosmetically and socially acceptable than a fabric mask.
- Very hot, does not breathe, and can cause maceration of the skin and even heat injury
- Poor compliance is common.

COMPRESSION GARMENTS

- Compression garments are used to help decrease hypertrophic scarring. It is thought that the pressure (at least 25 mmHg) exerted on capillaries reduces blood flow, thereby reducing scar formation.
- They are worn 23 hours per day until scars mature, which can be up to 2 years, and removed only for washing (Schneider & Spires, 2010). However, compression garments are often uncomfortable and difficult to put on, which often leads to poor compliance (Esselman, 2007). Up to 40% of adult burn patients are nonadherent (Crofton et al., 2020).
- They are specifically measured to fit the burned areas: Face masks, gloves, sleeves, jackets, or pants. Proper fit prevents friction from the garment to create superficial wounds in the burn scar (Gabriel & Holavanahalli, 2016).
- They will need to be replaced frequently, as they wear out and must be changed as the patient gains or loses weight.
- The location of the use of compression garments in treatment for severe facial and upper body burn is important. Obstructive sleep apnea (OSA) has been documented in cases with compression garment treatment. Therefore, extreme caution should be used in patients who have a history of OSA or are at risk for development (Schneider & Spires, 2010).
- Compression wraps are recommended to decrease hand edema, increase ROM, and increase hand function in patients with hand burns (Edger-Lacoursière et al., 2023).
- In children, the use of compression garments to treat hypertrophic scarring of the neck and head has been shown to cause deformities. Caution must be used in the application of pressure garments around the face, as they will impact facial growth and development of normal contours (Schneider & Spires, 2010).
- The use of pressure garments at the mandible may cause hypoplasia (Schneider & Spires, 2010).
- Poor compliance

Other Mechanisms to Control Hypertrophic Scarring and Contractures

- Silastic gel has been found to reduce hypertrophic scar formation in the absence of pressure. However, the mechanism of action is not clear. It is used to line silicone masks and is applied over individual problem scar areas.
- Steroids injected locally may reduce hypertrophic scarring.
- Early mobilization and splinting are encouraged to maintain ROM and prevent contractures.
- Use assistive devices as needed for ambulation, including AFOs.
- Lasers are being used for scar mitigation. Pulse dye lasers are used to decrease erythema. Fractional CO_2 lasers with increased power and penetration cut the scar, allowing it to grow back in a less linear, more random pattern of connections, improving mobility and decreasing volume. This does not replace compression but enhances the outcome of scar profile.
- Scar massage may be helpful in improving pain and pruritus in hypertrophic scarring in the short term (Ault et al., 2018).

Pain Control

- Pain can be severe from burns, and adequate pain control is needed. Long-acting narcotics are used for baseline or background pain, with short-acting medication for episodes of breakthrough pain. Procedural pain is treated with local, conscious sedation, or general anesthesia as indicated. Doses may be high, but better outcomes in healing, anxiety, and

rehabilitation are linked to good pain control. Patient-controlled analgesia may be used to help the patient feel in more control.
- Consideration must be given to other causes of discomfort (e.g., neuropathic pain, myofascial pain, pruritus, sleep deprivation, and anxiety). These conditions should be treated aggressively and specifically. Mobilization must not be limited by pain.
- The use of gabapentin has been shown to decrease pain perception and decrease overall narcotic requirements in burn patients.
- Addition of duloxetine may increase efficacy of other analgesics (Najafi et al., 2019).
- Pain management is a dynamic process. As healing and mobility improve, opportunities to decrease opioids must be taken to prevent opioid-induced hyperalgesia from becoming a complication.
- Use of virtual reality in rehabilitation has been shown to reduce pain (Lan et al., 2023).

Psychological Issues and Treatments for Burn Survivors

PSYCHOLOGICAL ISSUES

- Posttraumatic stress disorder (PTSD): 20% to 45% of patients may meet the criteria for diagnosis 1-year postinjury, requiring long-term follow-up of patients' psychological status (Esselman, 2007).
- PTSD and depression are predictors of social interactions and social activities at 2 years postinjury (Deng et al., 2024).
- Severe loss of personal identity with change in appearance
- Loss and change in position in the family and the community, loss of employment
- Financial and social stress
- Stress of constant pain
- Survivor guilt
- Depression
- Preexisting psychological disorders
- Adjustment disorders
- Older age, mental health diagnoses, and unemployment are associated with decreased satisfaction with life scores in patients with burn injury.

TREATMENT

- Antidepressants
- Counseling
- Peer support of other burn survivors (e.g., Phoenix Society)
- Early reintegration into the community; early and repeated emphasis upon return to vocational and avocational activities
- Music therapy

Nutrition

- Adequate calories must be given to maintain a positive nitrogen balance and promote muscle and skin repair. Healing will not take place without it, as the body is in a highly catabolic state.
- This may require 25 kcal/kg in addition to 40 kcal per 1% total BSA burn per day and 15 g of nitrogen per square meter of BSA per day (Gabriel & Holavanahalli, 2016).
- Additional vitamin C, vitamin A, zinc, copper, and manganese are important in wound care. The use of additional supplementation is essential.
- Malnutrition in burn patients prolongs length of stay in acute care (Caldis-Coutris et al., 2022).
- Once the acute phase has passed, another problem may appear.
- Overeating will cause weight gain in any area that has not lost fat cells and can lead to disfigurement secondary to relative obesity. Burned fat cells are not replaced.

Exercise

- Use of anabolic agents and exercise in individuals with severe burns can increase strength and lean body mass (Esselman, 2007).
- Active ROM
- Cardiovascular fitness
- Slow sustained stretching of the skin contractures:
  - Gentle application of superficial heat prior to stretch
  - Manual
  - Traction/weights
  - Serial casting/splinting
  - Paraffin
  - Massage/vibration

OTHER COMPLICATIONS

Peripheral Neuropathy

- Present in 15% to 20% of burn patients with BSA of 20% or greater (Helm et al., 1998)
- Burns of BSA of >20%, high-voltage electrical injuries, age > 40 years, and prolonged hospitalization more easily develop compression neuropathies (Tu et al., 2017).
- Can occur in burned or unburned areas
- In acute burn, may require nerve decompression, transposition, anastomosis, or nerve graft
- In delayed neuropathy, can be diagnosed by nerve conduction study (NCS)/EMG. Causes include entrapment, neuroma, scarring, HO
- May have paresthesias and/or weakness
- Can present as mononeuropathy, multiple mononeuropathies, or a generalized peripheral neuropathy

Bone and Joint Changes

- Osteoporosis is the most common finding.
- Decreased growth in children with burns near the epiphyseal plate. As a result, bone growth deformity can occur after burn injury.
- Also, must change the size of the compression garments frequently as children grow to prevent deformity. Particularly true with pressure to the mandible, which causes overbite.

Osteophytosis

- Found at the elbow and olecranon or coracoid process after burn injury

Heterotopic Ossification

- Ectopic bone deposition in periarticular soft tissue
- 🕮 In adults, the most common site of HO joint involvement in burns is the elbow, then the shoulder.
- In children, the most common site of HO joint involvement in burns is the elbow, then hips.
- Reported in up to 17% of burn patients
- Recommend pain-free active ROM

Amputations

- Complicated by fragile skin and contractures, resulting in difficulty with prosthetic fitting
- Major amputations related to high-voltage electrical injuries
- Finger amputations associated with thermal injuries

Scoliosis and Kyphosis

- Can be seen with burns of the chest or back
- May occur from protective posturing

Joint Subluxations and Dislocations

- Seen with burns of the dorsal surfaces of the hands and feet. During the healing process, skin pulls the joint into hyperextension. If this continues chronically, it can cause joint subluxation.
- Seen in MCP and metatarsophalangeal (MTP) joints
- Splint MCP joint in flexion between 60 and 90 degrees, and exercise
- Use a surgical high-top shoe with a metatarsal bar for 24 hours per day to prevent MTP subluxation.

🕮 Infections

- It is important to monitor for potential signs of burn wound infection. It is also important to note that while all burns are colonized with bacteria, not all burns are infected.
- Pyrexia and leukocytosis are commonly seen in burn patients without an active infection.
- Methicillin-resistant *Staphylococcus aureus* furunculitis: Hard, indurated boils that are painful with minimal purulence involving burned or nonburned areas.
  - Incision and drainage may not be sufficient for multiple furuncles and may require antibiotic treatment (trimethoprim/sulfamethoxazole) or multiple antibiotics.

POSTACUTE PHASE BURN CARE

- Continue local wound care.
- Prevention of new injury from mechanical irritants; the skin is now very sensitive.
- Lubricate skin several times a day.
- Compression garments should provide at least 25 mmHg or more pressure and be worn 23 hours per day to reduce hypertrophic scarring (Helm et al., 1998).
- Pruritus can be intense and difficult to treat. It is felt to be a component of neuropathic pain and not related to histamine release. Good results are obtained using gabapentin for pruritus.
- Provide protection from the sun as skin is susceptible to repeat burns. Wear long sleeves, hats, and sunscreen.
- The skin will also be susceptible to topical irritants, such as oil and gas, which should be avoided.
- Extreme heat should be avoided in full thickness burns, as the ability to cool the body through sweating is lost due to the loss of sweat glands. The inability to thermoregulate can limit physical activities and return to work in hot environments (Esselman, 2007).

BIOSTATISTICS

Measurements describe phenomena of interest in terms that can be analyzed statistically. The choice of the measurement scale is important, as it directly influences the information content of the measurement and enables different types of statistical analyses.

MEASUREMENT SCALES

- There are two main types of data:
  - Categorical (qualitative) data
  - Numerical (quantitative) data
- **Categorical data:** Phenomena that cannot be quantified are measured by classifying them into distinct, qualitative categories.
  - Categorical data with two possible values are called *dichotomous* or *binary* or *binomial* (e.g., living/dead, treated/untreated).
  - A **nominal scale** organizes data in *unordered*, named categories with no quantifiable intervals between categories (e.g., blood type, race, hair color).

  - 🕮 An **ordinal scale** classifies data within various categories possessing a natural order but do not necessarily have quantifiable or equal intervals (e.g., Functional Independence Measure [FIM] Scale, pain severity, disease staging, fracture type).
  - Descriptive statistics for categorical data include frequencies, percentages, or proportions in each category, sometimes accompanied by standard errors (later in this section). Examples: Percent of patients who are female or the proportion of patients with a certain type of injury.
- **Numerical data:** Numerical data quantify the phenomena that are measurable and can be expressed in terms of quantity or count. It can be broken into two types: Continuous and discrete.
  - **Continuous data** are collected on characteristics presented on a continuum.
    - Examples: Body weight, ROM, systolic blood pressure (BP), serum high-density lipoprotein
  - **Discrete data** describe an **interval scale** that uses measurements with equal unit intervals between each category, but the zero point on the scale is arbitrary. Interval scales can have positive or negative values.
    - Example: Temperature measured on Fahrenheit scale
  - A **ratio scale** is similar to an interval scale in that the intervals between measurements are equal, but the zero point on the scale is a real absolute zero indicating the absence of the phenomena being measured. Because the intervals are equal and the zero point is fixed, these measurements can also be expressed as ratios.
    - Examples: Height, weight
  - Three most common descriptive statistics of central tendency in continuous measurement data include:
    1. **Mean:** Average value of the data set
    2. **Median:** 50th percentile value in a data set. Half of the values in a data set are above the median value, and the other half of the values are below it.
    3. **Mode:** Most common value in the data set. The terms *bimodal* (two peaks) or *trimodal* (three peaks) are used if there is more than one peak frequency.
    - In a symmetric, bell-shaped distribution, the mean, median, and mode would all be equal.
  - Three most common measures of variability for continuous measurement data include:
    1. **Range:** The difference between the minimum and maximum values
    2. **Standard deviation** (SD or σ): The average amount of variability in the data set, indicating how far each score is from the mean
    3. **Standard error** (SE): Expected error in estimating sample mean or proportion using a sample of a certain size. SE = Sample SD/Square root of the sample size

DISTRIBUTIONS

- A **distribution** is a convenient tool used to summarize a set of data. Many observations in medicine fall into patterns that match well-known statistical distributions. This is useful because the probabilities of extreme observations can be readily calculated from known population distributions using the measures of location and variability described earlier.
- The **normal distribution (Gaussian curve or bell curve)** is a symmetric, bell-shaped distribution that matches the empirical distribution of many continuous medical measures. Each distribution has a mean (μ) and variance (σ^2).
- A ***Z*-score** represents the number of SD away a value is from the μ, the *population* mean. 1 *Z*-score unit = 1 SD.
  - −1 Z to +1 Z (±1 SD from the mean in a normal distribution) includes 68% of the observations, which equates to 68% of the area under the bell curve. Remember that the area under the bell curve is proportional to the frequency or probability of the observations.
  - −2 Z to +2 Z (±2 SD) includes about 95% of the area under the curve.
  - −3 Z to +3 Z (±3 SD) includes about 99.7% of the area under the curve.

  - In clinical medicine, the Z-score is the number of SD above or below a population mean (an "average person" of the same age, race, and gender).
- A ***t*-score** is the number of SD a *sample value* is above or below the mean in a sample distribution. (When the sample is very large, the t and Z distribution values are the same, so occasionally you will see them used interchangeably in publications.)

STATISTICAL TESTING

- A **statistical test** is a way to make conclusions or decisions concerning characteristics or outcomes in a larger patient population based on sample data. There are many forms of statistical testing, and each involves estimating the probability that an outcome could be observed as a chance fluctuation.
- **Possibility versus Probability**
  - **Possibility** is the potential that a particular event could occur. Note that an outcome can be possible but not probable. The concept of possibility is a subjective term and not a mathematical value.
  - **Probability** is the chance or likelihood of a particular event (possibility) occurring from a possible set of outcomes, typically expressed on a scale from 0 to 1 (or a percentage from 0% to 100%). A rare event has a probability close to 0 (0%), and a very likely event has a probability close to 1 (100%).
  - Example: The possibilities for a coin flip are landing on heads or on tails. The probability of landing on heads is 50% and on tails is 50%.
- **The null hypothesis (H_0)** represents a hypothesis/assumption that suggests no effect or no difference. A researcher hopes to disprove (nullify) H_0 to support an alternative hypothesis using statistical tests. It is typically the *opposite* to what the researcher is trying to prove.
  - Example: If a researcher is trying to prove that "Drug A" lowers BP better than the placebo, the null hypothesis would be that Drug A does not lower BP any more than placebo.
- A **Type I error (false-positive)** is the probability that the null hypothesis is rejected when it is actually true. The probability of committing a Type I error is called α (alpha). α is preset at an arbitrary level by the researcher. Conventional α levels are 0.05 (5%), 0.01 (1%), and 0.001 (0.1%).
- A **Type II error (false-negative)** is the probability of failing to reject the null hypothesis when it is false. The probability of making a Type II error is called β (beta). The likelihood of a Type II error depends on the magnitude of the difference measured within the data set and the sample size. If there is an inadequate sample size, a difference may remain undetected in a statistical comparison.
- **Statistical power (1 – β)** is the probability of correctly rejecting a false null hypothesis. Power is the chance of not making a Type II error. Several factors such as sample size, effect size, the variability of the data, and the type of test influence the statistical power.
- Statistical tests usually generate a ***p*-value,** which is the probability that a set of observations/results would be produced by random chance alone (i.e., due to a fluke of sampling).
  - 🕮 If the *p*-value is smaller than the alpha level (α), the results are considered statistically significant. Example: If the *p*-value was 0.001, the results would be considered statistically significant if you were using an α of 0.01 or 0.05, but not at 0.001.
- The **sample size** (n) should be published along with the results of any statistical analysis. This is different from **degrees of freedom** ("df"), which refers to a measure of the number of values that are allowed to vary and assume any value in a statistic.
- A **confidence interval (CI)** quantifies the precision of measurements obtained from a sample. A CI represents the probability that a value falls within an interval, signifying that the hypothesis is true.
  - In a normal distribution, the mean value is at the center of the CI and contained within symmetric upper and lower bounds of the CI. Distributions, such as *Z*- or *t*-distributions, are used to set the bounds of the CI.

- Typically, 95% CI ($\alpha = 0.05$) or 99% CIs ($\alpha = 0.01$) are utilized.
- If a certain specified value lies outside a 95% CI on a sample mean, then the sample mean is statistically significantly different at the 5% level ($\alpha = 0.05$), from that specified value.
- **Figure 12–13** is a visual display of 95% CIs for the number of venous thromboembolisms diagnosed according to the day of rehabilitation hospitalization. The CI for the "1–5" category does not overlap the others. Thus, it is statistically significantly different from the others with an alpha of 0.05 and a 95% CI. The CIs for the other categories all overlap.
  - Overlapping CIs lead to a conclusion of "no difference" (*not* statistically significantly different from the others).

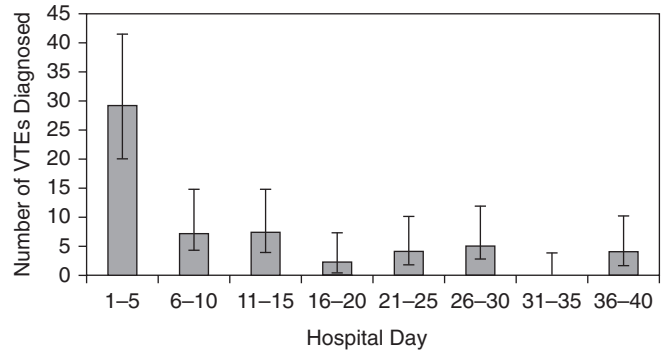


FIGURE 12–13 Frequency of venous thromboembolism by day of rehabilitation; an example of confidence intervals (CI). Lines on the bars indicate a 95% CI.
VTEs, venous thromboembolic diseases.

STUDY DESIGN

Various possible study designs are employed in the collection of medical data. Common types of study designs include:

- **Observational studies:**
  - Cohort (longitudinal) studies
  - Case–control studies
  - Cross-sectional studies
- **Interventional studies:**
  - Randomized controlled clinical trials
  - Nonrandomized clinical trials
- A **cohort study** (or **longitudinal study**) involves a patient population defined by initial exposure status/diagnosis or receiving a specific treatment and compared with another group of patients who are not subjected to the same exposure or treatment (e.g., radiation exposure vs. no radiation exposure). The subgroups are then followed over time to determine the outcomes in each. Comparisons may be made among subgroups or with untreated controls. Cohort studies can be prospective or retrospective studies. They usually require larger sample sizes.
  - Cohort Study Analysis:
    - **Absolute risk (incidence rate)** is the rate at which an outcome occurs per time interval.
    - **Risk ratio (RR),** also known as **relative risk,** compares the probabilities of developing an outcome between exposed and unexposed subgroups.
    - Example: RR = Probability of developing cancer after radiation exposure/probability of developing cancer with no radiation exposure.
    - RR = 0.50 means the relative risk is 50% less in the first group.
    - RR = 1 means there is no relation to exposure status and outcome because both subgroups have equal odds of developing the outcome.
    - RR = 1.5 means the relative risk in the first group is increased by 50%.

- A **case–control study** involves comparing a group of patients diagnosed with a disease/condition or outcome with a control group lacking it. Exposure histories are then compared between these groups. Case–control studies are typically (but not always) retrospective in nature, utilize smaller sample sizes, and are especially valuable in investigating rare diseases/outcomes. Case–control outcomes include odds ratios.
  - **Odds ratio (exposure odds ratio)** is the odds of case exposure/odds of control exposure. It is similar in interpretation to relative risk used in cohort studies.
    - Example: A person with a grade 3 or 4 pressure ulcer has a mortality odds ratio of 1.74, which means that a person with this condition has an increased mortality risk of 74%.

📖 *Note the contrast: Cohort studies follow a defined exposure group over time and provide estimated risks and relative risks as well as incidence rates. Case–control studies use data from a sample with disease or other endpoint, compare it with a control group in terms of exposure histories, and generate odds ratios. Risks or incidence rates are not available for case–control studies because cases and controls are not drawn from a definable denominator.*

- A **cross-sectional study (cross-sectional analysis)** represents a snapshot of data at one point in time. Cross-sectional studies provide **prevalence rates,** that is, the proportions/percentages of various characteristics, or distribution patterns, and average values of various variables for the patient population, but cannot provide causation and effect relationships.
  - Example: U.S. Census or the National Health and Nutrition Examination Survey, providing information about the health and habits of the U.S. population, such as the prevalence of smoking in various demographic groups.
- A **clinical trial** is a type of cohort study with the following characteristics:
  - It is **experimental** (i.e., interventional rather than purely observational).
  - It is conducted to determine the best course of therapy for future patients with a particular condition based on the responses in a sample.
  - Assessment of outcome is often conducted in a "**double blinded**" fashion. Double blinding a study means both the patient and the evaluator are unaware of the type of treatment the patient receives. This is to avoid **a biased assessment** of the outcome.
  - Patients are assigned to either a treatment group or a control group. Controls may receive placebo therapy or an existing standard of care.
  - Patient assignment is typically randomized to avoid **sample bias,** which could distort the outcome of that treatment. Examples of bias include using only the healthiest patients to evaluate the response to a specific treatment or confounding by indication (the anticipated optimal treatment is selected for each patient).
- A **case series** is a group of **case reports** collected under widely varying conditions and usually without controls. They may use only descriptive statistics or no statistics at all. Denominators are not available, so rates cannot be calculated.
  - Sometimes called *anecdotal reports*, which may have a pejorative connotation.
  - Are useful for publicizing unanticipated possibilities, sentinel events, or novel preliminary findings of clinical interest.

Other Key Definitions and Elements of Study Design

- **Prevalence:** Total number of people with a condition in the population at one point in time. **Prevalence rate** is the percentage of cases found in the population with the condition.
- **Incidence:** Number of new cases of a condition or outcome during the follow-up period. **Incidence rate** is the percentage of new cases of a condition or outcome that have occurred in the population.
- **Inclusion criteria:** A set of attributes/characteristics required of the potential subjects to be considered eligible for a study.
- **Effect size:** A measure of a difference or size of association the investigator wishes to detect between the groups.
- A **prospective study** is a study that looks forward in time in relation to an outcome. Clinical trials and most (but not all) cohort studies are considered prospective studies.

- A **retrospective study** looks backward in time to an event that has already occurred. Case reports/case series and most (but not all) case–control studies are considered retrospective studies.
- **Sample selection** should represent the group the items are selected from (i.e., unbiased, independently, and randomly selected) and large enough to provide reliable estimates of measurements (i.e., "*adequately powered*" to avoid Type II errors).
- ***t-test* (or Student's *t-test*):** A statistical tool used to determine whether the mean value of a variable in one group differs significantly from that in another group. A *t-test* can be used to compare two independent groups (independent samples *t-test*) or compare dependent groups (paired samples *t-test*) such as before-and-after comparison of the same group.
- **Reliability** refers to the reproducibility of a study.
- **Validity** describes the extent to which a study accurately measures what it sets out to through its study design and methods. Validity can be affected by multiple factors, including systematic errors in the study such as bias. While some studies are more prone to bias, all studies have some degree of bias.
  - **Bias** refers to distortion of data from the methodology of data collection or patient selection.
  - Common biases include the following:
    - **Selection bias** involves picking or using patients that favor a particular outcome.
    - **Sampling bias** refers to prejudice toward or against a certain outcome due to an incomplete/insufficient representation of the population.
- Approval of an Institutional Review Board is a legal requirement that must be fulfilled prior to conducting any research involving human subjects.

ASSESSMENT OF SCREENING AND DIAGNOSTIC TESTS

- A screening or diagnostic test may be faster, less expensive, or less invasive than a "gold standard" test, but it is important to make sure that it yields comparable results. The main measures of comparability of tests are **sensitivity** and **specificity.**
- **Sensitivity** is a measure of how often a test will detect the *presence* of the clinical problem in a group of patients known to have it.
- **Specificity** is a measure of how often a test will indicate the *absence* of the clinical problem in a group of patients who are known *not* to have it.
- **Positive predictive value (PPV)** measures the probability that a positive test is truly positive.
- **Negative predictive value (NPV)** measures the probability that a negative test is truly negative.

TEST OUTCOME	DISEASE	NO DISEASE	PREDICTIVE VALUE
Positive test	a (True positive)	b (False positive) Type I error	PPV = a/(a + b)
Negative test	c (False negative) Type II error	d (True negative)	NPV = d/(c + d)
	Sensitivity = a/(a + c)	Specificity = d/(b + d)	

NPV, negative predictive value; PPV, positive predictive value.

BASIC PRINCIPLES OF CLINICAL ETHICS

Clinical ethics utilizes a structured approach to guide ethical decisions in clinical medicine. Bioethics scholars generally identify four ethical principles that are relevant to clinical medicine: Beneficence, nonmaleficence, justice, and respect for autonomy.

Autonomy

- **Autonomy** (*autos* = self and *nomos* = governance) indicates self-determination. In healthcare, it refers to one's moral right to make knowledgeable decisions about one's own care.
- Autonomy assumes that one has the capacity to make a decision, understands all the relevant information, and has the power to act on the decision without constraints.
- Respecting a patient's autonomy may include asking them if they wish to be the decision-maker, have family members assume this role, or enact a combination of both.
  - A common conflict in clinical ethics is that of autonomy versus **paternalism.**
    - In the *paternalistic approach*, the clinician decides what is best for the patient. In cases where patients prefer not to be involved in decision-making, or in emergency cases where physicians act based on what they think a patient needs, a paternalistic model may be appropriate.
    - In nonemergent situations, the contractual model, which recognizes the right patients have to make informed choices about their treatment, should be used.

🕮 Beneficence

- Refers to an action that *benefits* the patient.
- Example of beneficence: A speech therapist recommends the use of a percutaneous endoscopic gastrostomy (PEG) tube as the most appropriate treatment for satisfactory nutritional intake in a patient with persistent, severe dysphagia. If the patient declines and chooses to continue oral intake, the principle of autonomy is being exercised.

Nonmaleficence

- Refers to a central tenet to medical ethics and means "do no harm." It is derived from the Latin expression *Primum non nocere* ("First, do not harm.")
- In professional ethics, not harming a patient is a separate principle from actions that benefit a patient (beneficence).
- In the dysphagia case described previously, while autonomy may conflict with beneficence, maleficence also arises because of the possibility of harm from the recommended intervention.

🕮 Justice

- In biomedical ethics, discussions on justice usually focus on the fair distribution of limited healthcare resources. The limited resource may be patient beds, healthcare professional services, medications, respirators, treatment modalities, or diagnostic tests and procedures.
- Justice implies fairness.
- **Comparative justice** refers to allocation of resources based on needs or wants.
  - Example: If vaccines have limited availability, give vaccinations to only high-risk patients.
- **Noncomparative justice** favors an equal share over individual needs.
  - Examples: Equal limitations on therapy services, prospective payment systems, and diagnosis-related groups.

Informed Consent

- Informed consent requires three elements. It is largely based on the principle of autonomy and requires (mnemonic "IVC"):
  - **I**nformation (disclosure)
  - **V**oluntary (no coercion)
  - **C**apacity (see below)

Capacity

- Capacity refers to a person's ability to make a decision regarding treatment.
- Capacity is decision-specific. This means that the patient may have capacity to decide whether or not to have blood drawn, but does not understand (and therefore does not have capacity to make a decision) complex surgery.

- Any physician can make a determination about capacity. If there are questions regarding capacity, a psychiatric consultation can be obtained.
- Capacity involves four factors, all of which the patient must demonstrate (mnemonic "CUAR"):
  - **C**ommunicate: The patient must be able to communicate their decision. Patients in the final stages of ALS may lose the ability to communicate. Therefore, it is important to document their healthcare wishes before the disease progresses.
  - **U**nderstand: The patient must understand the implications of accepting the treatment (and the consequences of refusing treatment).
  - **A**ppreciate: The patient must be able to appreciate the consequences of deciding on a treatment.
  - **R**ationale: The patient should be able to give a rationale for deciding or refusing a treatment.
- One helpful way to assess capacity would be for the patient to verbalize what they understand about the treatment, what would happen if they refuse a treatment, and explain why they are refusing a treatment. A patient's capacity should not be questioned just because they disagree with what a physician proposes.
- When a patient's decisional capacity is impaired, alternatives include:
  1. **Advance directive** prepared by the patient. This is a written document indicating what treatments the patient would or would not want (e.g., at the end of life, should only comfort care be given, or should all measures be taken to prolong life?). Advance directives include living wills, power of attorney for healthcare (medical power of attorney), physician orders for life-sustaining treatment), do not resuscitate (DNR) orders, and organ and tissue donation.
  2. Appointment of a **surrogate decision-maker** (if there is no advance directive). A surrogate decision-maker should always use substituted judgment (i.e., what would the **patient** want in these circumstances, not what would **I** want in these circumstances?).
     a. **Durable healthcare power of attorney** is a legally appointed proxy who can make healthcare decisions **if** the patient loses capacity.
     b. In some states, there is a legal hierarchy for choosing the appointed surrogate decision-maker if there is no written surrogate decision-maker (e.g., the New York Family Health Care Decisions Act allows family members or close friends to make healthcare decisions if there is no healthcare proxy and the patient has been determined to lack capacity). Physicians should always follow the laws of the state where they are practicing.
  3. If a patient does not have a designated proxy, alternatives include:
     a. Person designated by state surrogate statutes (usually a spouse, followed by adult children, parent, or close friend)
     b. Court-appointed guardian
     c. The patient's family, friends, and physician collaborating to select the most appropriate person

Competence

- Competency is a legal description in which only judges have the right to rule that a person is incompetent.

Moral Distress

- Moral distress occurs when providers believe that they are being involuntarily complicit in acting unethically. They are doing something that they believe to be morally wrong but have little power to act differently or to change the situation. In many ethical healthcare situations, the staff may feel moral distress when they believe decisions are incorrect or that the decision goes against their own moral belief.

Ethics Committee

- All hospitals that are Joint Commission-approved must have a mechanism for resolving ethical issues in healthcare. It recommends a multidisciplinary ethics committee. This committee should be composed of various healthcare providers, including clergy, social workers, nurses, and physicians. There should be a lay person from the community as well. According to the

American Medical Association (AMA) Medical Code of Ethics, the "Ethics Committees, or similar institutional mechanisms, offer assistance in addressing medical issues that arise in patient care and facilitate sound decision making that respects participants' values, concerns and interest" (AMA, 2017). The committee should serve as advisors and educators, rather than decision-makers.

Ethics Consultation

- Many hospitals have an ethics consultation service that performs ethics consults (similar to calling for a Physical Medicine and Rehabilitation [PM&R] consult). In most hospitals, anyone can call an ethics consult (physician, staff, patient, or family member). As with any consultation, the conclusions of the consultant are meant to be recommendations and not requirements. There is now certification for professionals who perform ethics consults (Healthcare Ethics Consultant-Certified). Although this is not a requirement, certification demonstrates a certain level of proficiency.

The Four-Quadrant Method for Ethical Dilemmas

- The four-quadrant method **(Table 12–12)** provides a way to organize the information of a clinical ethical problem into four main topics:
  1. **Medical indications** are the clinical data necessary to diagnose the extent and severity of medical problems, to make a prognosis for survival and restoration of impairments, and to determine the options for treatment, which include risks, benefits, and likely success of each treatment option. A clear view of the possible benefits of interventions is the first step in assessing the ethical aspects in this case.
  2. **Patient preferences** are the expressed choices of the patient or the surrogate decision-maker.
  3. **Quality of life** refers to medically pertinent features of the patient's life before and after treatment.
  4. **Contextual features** identify the familial, social, institutional, financial, and legal settings that influence medical decisions.

- Ethics case: Donald "Dax" Cowart is a survivor of a 1973 propane gas explosion which left him severely handicapped. He was 25 years old when he was severely burned over 65% of his body with severely damaged eyes. Initially, his survival was in doubt. He was eventually stabilized and underwent amputation of several fingers and extraction of his right eye. During much of his hospital and inpatient rehabilitation stays that altogether lasted for more than a year, he repeatedly demanded that he be allowed to die. Despite his demands to his mother, lawyer, and physicians, his treatments were continued. Upon discharge, he was completely blind, with minimal use of his hands, and dependent on others to assist in personal daily activities.

1. **Medical indications:** Dax's survival probability was approximately 20% after initial resuscitation and increased to almost 100% after 6 months of hospital care. Treatments such as fluid resuscitation, skin grafting surgeries, and antibiotics are associated with certain probabilities of success and risk.
2. The **patient's preferences** to initially reject care were overridden by his physicians due to doubt in his decisional capacity. However, a psychiatric consultation later confirmed his decision-making capacity was not impaired. Should Dax's preference have been respected? Did Dax appreciate fully the prospects of his rehabilitation? Should the doctors be obliged to pursue therapies they believe have promise over the objection of the patient? Would the doctors be assisting in a suicide if they followed Dax's wishes?
3. **Quality of life:** Dax was a popular, athletic man who was working in real estate. After the accident, he suffered from excruciating pain and profound depression. He also had significant physical handicaps and disabilities, including amputations, blindness, and limitations of daily living activities. Was the prospect for return to what is considered a normal or acceptable life so poor that no reasonable person would choose to live in such a way? Who would make such decisions and what values would be used in making such a decision?

TABLE 12–12 Four-Quadrant Method

MEDICAL INDICATIONS	PATIENT PREFERENCES
BENEFICENCE AND NONMALEFICENCE PRINCIPLES	**THE PRINCIPLE OF RESPECT FOR AUTONOMY**
• What is the patient's medical problem? History? Diagnosis? Prognosis? • Is the problem acute? Chronic? Critical? Emergent? Reversible? • What are the goals of treatment?	1. What does the patient want? What are the patient's goals? 2. Has the patient been informed of benefits and risks, understood, and given consent? 3. Is the patient mentally capable and legally competent? Is there evidence of incapacity?
• What are the probabilities of success of various treatment options? • What are the plans in case of therapeutic failure? • In sum, how can this patient be benefitted by medical and nursing care, and how can harm be avoided?	4. Has the patient expressed prior preferences (e.g., advance directives)? 5. If incapacitated, who is an appropriate surrogate? Is the surrogate using appropriate standards? 6. Is the patient consenting voluntarily? 7. In sum, is the patient's right to choose being respected to the extent possible in ethics and law?
QUALITY OF LIFE	**CONTEXTUAL FEATURES**
AUTONOMY, BENEFICENCE, AND NONMALEFICENCE PRINCIPLES	**THE PRINCIPLE OF JUSTICE**
1. What are the prospects, with or without treatment, for a return to the patient's normal life? 2. Are there biases that might prejudice the provider's evaluation of the patient's quality of life? 3. What physical, mental, and social deficits is the patient likely to experience even if treatment succeeds? 4. Is the patient's present or future condition such that continued life might be judged undesirable by them? 5. What are the plans and rationale to forgo treatment? 6. What are the plans for comfort and palliative care? 7. What ethical issues arise concerning improving or enhancing a patient's quality of life?	1. Are there family issues that might influence treatment decisions? 2. Are there provider (physician, nurses) issues that might influence treatment decisions? 3. Are there financial or economic factors? 4. Are there religious or cultural factors? 5. Is there any justification to breach confidentiality? 6. Are there problems of allocation of scarce resources? 7. What are the legal implications of treatment decisions? 8. Are there considerations of clinical research or teaching that might affect clinical decisions? 9. Any provider or institutional conflict of interests? 10. Are there public health and safety issues that affect clinical decisions?

Source: From Jonsen AR, Siegler M, Winslade WJ. *Clinical Ethics: A Practical Approach to Ethical Decisions in Clinical Medicine*. McGraw Hill; 2002, with permission.

4. **Contextual features** in this case included the legal implications of honoring Dax's wishes, Dax's mother's opposition to cessation of medical treatment due to religious reasons, and the massive cost of several months of hospitalization.

Although Dax views his life differently now, he continues to believe that he should not have been deprived of the right to end his life.

MULTIPLE SCLEROSIS

- MS is the most common demyelinating disease of the CNS and is the most common cause of nontraumatic disability in young adults. It is characterized by focal demyelination with relative axonal sparing due to immune-mediated inflammation. These changes result in the formation

of hallmark white matter plaques in the brain and spinal cord that may recur and/or enlarge with subsequent exacerbations.
- An immune-mediated response causes axonal degeneration with loss of oligodendrocytes, loss of myelin sheaths, and demyelination. Plaque formation results in a neurodysfunction of conduction (partial or complete), making action potential propagation down the nerve impossible. Older plaques may become sclerosed due to oligodendrocyte destruction, astrocyte proliferation, and glial scarring. In acute lesions, remyelination may occur, which results in a "remission."

FACTS

- MS is the third leading cause of significant disability in the 20- to 50-year age range.
- Mean onset is age 30.
- Women account for approximately 80% of cases; it affects Caucasians >>> African Americans, Asians (middle-aged adults), and women of northern European descent of childbearing age.
- The exact cause is unknown.
- Theories of pathogenesis:
  - Immune-mediated inflammatory process resulting in neurodegeneration
  - Genetic factors: Family history (siblings > children); increased risk associated with some Human leukocyte antigens-A and other risk loci have been found. Twins: Dizygotic same as siblings at 3% to 5%. Monozygotic 25% risk.
  - Immunologic: T-cells travel to the inflamed areas crossing the blood–brain barrier.
  - Viral: Slow virus—no specific evidence.
  - Environmental triggers may play a role in the development of MS in a person of genetic susceptibility, but further research is needed (Ghasemi et al., 2017).
- Geographic distribution: Prevalence rate is <1 per 100,000 in equatorial area compared with 4 to 6 per 100,000 in southern Europe and southern United States. In Canada, northern Europe, and northern United States, the rate is 30 to 80 cases per 100,000. It is estimated that there are nearly one million people in the United States with MS.
- Migration to temperate climate before adolescence
- 🕮 Pregnancy decreases relapses but then increases after delivery. No change in long-term prognosis.
- Potential role of vitamin D: Low vitamin D level is a risk factor for development of MS and also modifies disease activity.
- Cigarette smoking is a risk factor.

THE FOUR MAJOR PATTERNS OF MULTIPLE SCLEROSIS AS DESCRIBED BY THE NATIONAL MULTIPLE SCLEROSIS SOCIETY

1. Clinically Isolated Syndrome

- A first attack compatible with MS, with evidence of inflammation and demyelination
- Typically lasts a minimum of 24 hours
- Does not yet meet the criteria for MS

2. Relapsing-Remitting Multiple Sclerosis

- 🕮 The most common pattern: 85% to 90% begin with relapsing-remitting multiple sclerosis (RRMS): Clearly defined exacerbations of new or increasing symptoms, followed by partial or complete recovery or remission.
- Pattern: Early exacerbations followed by a complete remission. There are long periods of stability, with a return to baseline or mild disability after each exacerbation. Characterized as active (with relapses) or not active; worsening or not worsening.
- Gender-dependent, female > male by 2:1 ratio

3. Secondary Progressive Multiple Sclerosis

- RRMS that converts to steady deterioration with or without relapses; characterized as active (with relapses) or not active with progression or without progression
- Also gender-dependent, female > male by 2:1 ratio
- 50% of RRMS convert to secondary progressive multiple sclerosis (SPMS) after 10 years; 90% after 25 years

4. Primary Progressive Multiple Sclerosis

- Worsening as soon as symptoms appear: An insidious onset with a steady progression of symptoms, few remissions, and increasing disability. A progression to death can occur in weeks to months. It is more common in the older population, and male to female ratio is 1:1. Characterized as active (occasional relapses or new MRI activity) or not active, with progression or without progression
- 10% of cases at disease onset
- Worse prognosis

OTHER PATTERNS OF MULTIPLE SCLEROSIS INCLUDE:

5. Benign Multiple Sclerosis

- Functional 15 years after onset
- This results in mild symptoms, early exacerbations, and complete remissions (resolution of inflammation, partial remyelination, or rerouting of nerve transmission) with minimal or no disability seen after remission.

6. Malignant

- Rapid course, severe, in short period of time

🕮 PROGNOSTIC FACTORS OF MULTIPLE SCLEROSIS (TABLE 12–13)

TABLE 12–13 **Prognostic Factors of MS**

FACTORS	GOOD PROGNOSIS	POORER PROGNOSIS
Symptoms	Monosymptomatic	Polysymptomatic
Onset	Sudden, good recovery with long remission	Rapidly progressive
Findings at onset	Sensory optic neuritis	Motor (first sign) ataxia and tremor
Ambulation	Yes	No
Remission and relapses	Longer, more complete remissions	Higher relapse rate
Disability	Low current disability	High disability

MS, multiple sclerosis.

SIGNS AND SYMPTOMS OF MULTIPLE SCLEROSIS

Signs and symptoms of MS will depend on the location of the white matter lesion in the CNS. The onset of the disease may often present as optic neuritis or transverse myelitis. In advanced stages, the Charcot triad (scanning speech, intention tremor, and nystagmus) may be seen. Paresthesias and gait disturbances are commonly seen as an initial presentation. However, the most common clinical picture is a mixture of the abnormalities listed in the text that follows.

🕮 Common Symptoms

- Bladder, bowel, and sexual dysfunction
- 🕮 Fatigue (central in nature), reported in 90%

- Pain: Dysesthetic, "MS hug"; spasticity: painful spasms common
- Visual disturbances: Optic neuritis, diplopia, nystagmus, internuclear ophthalmoplegia
- Heat sensitivity (Uhthoff phenomenon)
- Cognitive impairment: Memory, attention, processing speed
- Cerebellum and basal ganglia: Ataxia, intention tremor, gait dysfunction, slurred speech
- Dorsal columns: Sensory abnormalities, including paresthesias, impairment of deep sensation, and proprioception
- Corticospinal tract: Weakness and spasticity
- Frontal lobe dysfunction: Cognitive deficits, memory, learning, impaired emotional responses, depression
- Speech abnormalities: Dysarthria, dysphonia
- Brainstem abnormalities: Myokymia, deafness, tinnitus, vertigo, vomiting, transient facial anesthesia, dysphagia
- Epilepsy
- Sleep disorders, OSA, RLS
- Vertigo, dizziness, imbalance
- Sexual dysfunction
- Note: Not all new symptoms result from a new MS lesion. Temporary aggravation of symptoms in old and previously silent lesion(s) may be caused by fever, heat, stress, fatigue, or other medical problems, especially a pulmonary or urinary tract infection, dehydration, or medication side effects. Aggravating factors and other medical problems must either be identified and treated or ruled out.

Top Three Problems Affecting ADLs Reported by Patients

1. Fatigue
2. Balance difficulties
3. Weakness

ADLs, activities of daily living.
Source: From Kraft GH, Freal JE, Coryell JK. Disability, disease duration, and rehabilitation service needs in multiple sclerosis: patient perspectives. *Arch Phys Med Rehabil*. 1986;67:164–168. doi:10.1016/0003-9993(86)90060-2.

🕮 Common Signs (Multiple Sclerosis Society of Canada, 2019)

- **Lhermitte's sign:** This is classically seen in MS but *not* pathognomonic for the disease.
  - Passive neck flexion causing an electric shock-like sensation radiating to the spine and shoulders, as well as other areas. This sign is most likely a result of the increased sensitivity of the myelin to stretch or traction.
- Upper motor neuron signs: Hyperreflexia, positive Hoffman and Babinski responses, spasticity
- Weakness
- Decreased sensation

DIAGNOSIS OF MULTIPLE SCLEROSIS

Clinical Criteria

- Diagnosis requires a history, examination, and exclusion of non-MS etiologies, and requires evidence of two separate attacks and damage to two separate areas of the CNS (involvement of two or more areas of CNS at different points in time).
- No single test is pathognomonic.
- CIS, first episode of neurologic symptoms typical of MS relapse in person not known to have MS:
- The 2017 Revised McDonald criteria **(Table 12–14)** combines clinical and neuroimaging elements in the diagnosis of MS. The 2017 revision allows other findings to support dissemination such as CSF oligoclonal bands, both symptomatic and asymptomatic MRI lesions, and cortical lesions.
- The criteria should be applied only to patients with typical demyelinating syndrome, to decrease the risk of misdiagnosis of MS (Solomon et al., 2018).
- To avoid misdiagnosis of MS: If not typical CIS, additional testing may be advised.

TABLE 12–14 2017 Revised McDonald MS Diagnostic Criteria

CLINICAL PRESENTATION	ADDITIONAL DATA NEEDED TO MAKE MS DIAGNOSIS
. . . in a person with a typical attack/CIS at onset	
• ≥2 attacks and objective clinical evidence of ≥2 lesions • ≥2 attacks and objective clinical evidence of 1 lesion with historical evidence of prior attack involving lesion in different location	None. DIS and DIT have been met.
• ≥2 attacks and objective clinical evidence of 1 lesion	**One** of these criteria: • **DIS:** Additional clinical attack implicating different CNS site • **DIS:** ≥1 **symptomatic or asymptomatic** MS-typical T2 lesions in ≥2 areas of CNS: Periventricular, **juxtacortical/cortical,** infratentorial, or spinal cord
• 1 attack and objective clinical evidence of ≥2 lesions	**One** of these criteria: • **DIT:** Additional clinical attack • **DIT:** Simultaneous presence of both enhancing and nonenhancing **symptomatic or asymptomatic** MS-typical MRI lesions • **DIT:** New T2 or enhancing MRI lesion compared with baseline scan (without regard to timing of baseline scan) • **CSF-specific (i.e., not in serum) oligoclonal bands**
• 1 attack and objective clinical evidence of 1 lesion	**One** of these criteria: • **DIS:** Additional attack implicating different CNS site • **DIS:** ≥1 MS-typical **symptomatic or asymptomatic** T2 lesions in ≥2 areas of CNS: Periventricular, **juxtacortical/cortical,** infratentorial, or spinal cord **AND** **One** of these criteria: • **DIT:** Additional clinical attack • **DIT:** Simultaneous presence of both enhancing and nonenhancing **symptomatic or asymptomatic** MS-typical MRI lesions • **DIT:** New T2 or enhancing MRI lesion compared with baseline scan (without regard to timing of baseline scan) • **CSF-specific (i.e., not in serum) oligoclonal bands**
. . . in a person with progression of disability from onset	
• Progression from onset	• 1 year of disability progression (retrospective or prospective) **AND** **Two** of these criteria: • ≥1 **symptomatic or asymptomatic** MS-typical T2 lesions (periventricular, **juxtacortical/cortical,** or infratentorial) • ≥2 T2 spinal cord lesions • CSF-specific (i.e., not in serum) oligoclonal bands

Note: Diagnosis of MS requires elimination of more likely diagnoses and demonstration of dissemination of lesions in the CNS in space and time. See source for details.
CIS, clinically isolated syndrome; CNS, central nervous system; CSF, cerebrospinal fluid; DIS, dissemination in space; DIT, dissemination in time; MS, multiple sclerosis; T2 lesion, hyperintense lesion on T2-weighted MRI.
Source: Adapted from Thompson AJ, Banwell BL, Barkhof F, et al. Diagnosis of multiple sclerosis: 2017 revisions of the McDonald criteria. *Lancet Neurol*. 2018;17(2):162-173. doi:10.1016/S1474-4422(17)30470-2.

Clinical Findings

- 🕮 Lesions scattered in time and space based on clinical findings or a clinical finding with MRI finding
- Space: By MRI in at least two of four MS typical regions of CNS or by clinical attack which involves a different CNS site

- Time: A second clinical attack or a new T_2 and/or gadolinium enhancing lesion
- Time between attacks: 30 days
- Two or more attacks with clinical evidence of two or more lesions
- Neurologic deficits in two or more areas, reflecting white matter involvement, at two points in time for >24 hours separately by 1 month
- Age of onset: 15 to 50 years old
- Typically two separate lesions in which the symptoms cannot be explained by a single lesion
- Objective deficits seen on exam
- Feature of typical signs and symptoms supported by diagnostic data

Diagnostic Studies

- 🕮 There is no pathognomonic test for MS. All test results are considered nonspecific and are to be interpreted within the clinical picture.

CEREBROSPINAL FLUID EXAMINATION

- Not required but can support diagnosis when clinical and MRI evidence is insufficient. Can substitute for demonstration of dissemination of lesions in time in some settings.
- Oligoclonal IgG bands (OCB) is the most important CSF test found in >95% with clinically definite MS and 60% to 70% of CIS. Presence is not sufficient for diagnosis. Presence in mono-symptomatic patients predicts higher rate of progression to MS.
- IgG index: Abnormal CSF IgG production found in 90% of clinically definite MS.

VISUAL EVOKED POTENTIALS (HIGH SENSITIVITY ALONG WITH MAGNETIC RESONANCE IMAGING)

- Abnormal (increased or delayed) P100 latency in 85% of clinically definitive MS and 30% with CIS

BRAINSTEM AUDITORY EVOKED RESPONSE

- Abnormal in 67% of clinically definitive MS
- Investigates the pontine area displaying an absence or delay of wave formation secondary to the demyelinating process.

SOMATOSENSORY EVOKED POTENTIALS

- Abnormal in 77% of clinically definitive MS
- In MS, the most frequent abnormality is latency increase or absence of component evoked by tibial nerve stimulation.

ELECTROMYOGRAPHY/NERVE CONDUCTION STUDY

- From a technical standpoint, standard NCS/EMG studies evaluate only the peripheral nervous system (PNS) and should not be affected since MS is a central process. However, there are cases of MS that affect the PNS on NCS/EMG.
- NCS assess PNS function and are typically normal (MS lesions affect CNS system). Decreased nerve waveform amplitudes may occur due to muscle atrophy.
- Needle EMG may show abnormal activity (e.g., fibs, positive sharp waves, facial myokymia) as can also be seen in other CNS lesions such as a stroke or TBI.
- Single-fiber EMG: Increased jitter (Grana & Kraft, 1994).
- The blink reflex tests central and peripheral nerve pathways and may be abnormal.

MAGNETIC RESONANCE IMAGING

- Test of choice to support clinical diagnosis of MS and has the greatest sensitivity. The McDonald criteria include MRI criteria.

- White matter lesions seen in 90% of clinically definitive MS and 80% of CIS who then develop MS.
- These ovoid-appearing plaques are typically located in the periventricular region, corpus callosum, centrum semiovale, or basal ganglia.
- Most sensitive indicator of disease activity. Monitors disease activity and progression.
- May visualize subclinical lesions: Enhancement with gadolinium may precede onset of deficits and can indicate active disease.

COMPUTED TOMOGRAPHY SCAN

- Not effective in visualizing lesions in the brainstem, cerebellum, and optic nerve
- Cerebral atrophy is the most common finding.

TREATMENT OF MULTIPLE SCLEROSIS

During an acute exacerbation, treatment should include a comprehensive rehabilitation program in addition to pharmacotherapy. Relative rest, hydration, dietary care (nutrition), bladder and bowel management, PT, occupational therapy (OT), and speech therapy (swallowing evaluations and protection, cognitive evaluations) are essential components of patient care.

Medications

CORTICOSTEROIDS (METHYLPREDNISOLONE)

- Used in short bursts for acute attacks secondary to its anti-inflammatory and antiedema effects. Acute attacks = "exacerbation," which is new or worsening MS symptoms lasting >24 hours and not related to metabolic factors (e.g., infection, stroke, renal failure).
- Dosing: 500 to 1,000 mg IV daily for 3 to 7 days with or without an oral taper, or oral prednisone can be considered.
- Oral is not used in patients with optic neuritis.
- Risks: GI disturbance, fluid retention, mood swings, electrolyte imbalance, insomnia, acne, hyperglycemia, hypertension
- Most responsive symptoms: Optic neuritis, brainstem, motor, acute pain, bowel, and bladder
- Least responsive symptoms: Cerebellar, sensory
- Long-term use leads to increased risk of long-term corticosteroid side effects: Hypertension, osteoporosis, diabetes, weight gain, and cataracts.
- Hastens recovery but does not prevent further attacks or alter disease progression

DISEASE-MODIFYING THERAPIES

- The following U.S. FDA-approved disease-modifying therapies for MS have been found through clinical trials to:
  1. Reduce the number of relapses
  2. Delay progression of disability
  3. Limit new disease activity (as seen on MRI)

INJECTABLE, ORAL, AND INFUSED MEDICATIONS

(A) Injectable Medications

1. **Interferon Beta:** Not for progressive MS in the absence of relapses; reduce relapse and disability progression
   - Interferon beta-1a: Used for CIS, RRMS, ASPMS (active secondary progressive MS- SPMS with relapses). Adverse effects include injection site reactions/necrosis, decreased peripheral blood counts, hepatic injury, depression, seizures, heart failure.
     - Avonex® (interferon beta-1a)
     - Plegridy® (peginterferon beta-1a)
     - Rebif® (interferon beta-1a)
   - Interferon beta-1b: Used for CIS, RRMS, ASPMS. Adverse effects include depression, drug-induced lupus, flu-like symptoms, hepatic injury, heart failure, and infection site reaction/necrosis.

- Betaseron® (interferon beta-1b)
- Extavia® (interferon beta-1b)

2. **Glatiramer:** Used for CIS, RRMS, ASPMS. Adverse effects include chest pain, hepatic injury, and postinjection reactions. Decreases relapse rate.
   - Copaxone® (glatiramer acetate)
   - Glatiramer Acetate Injection (glatiramer acetate—generic equivalent of Copaxone 20-mg and 40-mg doses)
   - Glatopa® (glatiramer acetate—generic equivalent of Copaxone 20-mg and 40-mg doses)
3. Monoclonal antibody
   - Kesimpta® (ofatumumab); monoclonal antibody used for CIS, RRMS, SPMS. Adverse effects include hepatitis B virus (HBV) reactivation, progressive multifocal leukoencephalopathy (PML), and infections

(B) Oral Medications: Decrease relapse rate, alter course of disease

1. For treatment of CIS, RRMS, active SPMS:
   - Aubagio® (teriflunomide): Adverse effects include bone marrow suppression, DRESS (drug reaction with eosinophilia and systemic symptoms), hepatotoxicity, hypertension, infection, malignancy, peripheral neuropathy, interstitial lung disease, Stevens–Johnson syndrome, and toxic epidermal necrosis.
   - Fumarates: Adverse effects include flushing, herpes zoster/opportunistic infection, hepatic injury, lymphopenia, and PML:
     - Dimethyl Fumarate (dimethyl fumarate—generic equivalent of Tecfidera)
     - Tecfidera® (dimethyl fumarate)
     - Vumerity® (diroximel fumarate)
     - Bafiertam™ (monomethyl fumarate)
   - Spingosine-1-phosphate receptor modulators: Adverse effects include bradyarrhythmia, hepatic injury, hypertension, infections including PML, macular edema, malignancies, adverse pulmonary function, and encephalopathy. Discontinuation of fingolimod may result in severe worsening of disease and permanent disability.
     - Galena® (fingolimod): First oral disease-modifying therapy approved for MS
     - Tascenso ODT® (fingolimod): Bioequivalent to Gilenya but not a generic product
     - Mayzent® (siponimod)
     - Ponvory™ (ponesimod)
     - Zeposia® (ozanimod)
2. For treatment of RRMS and SPMS due to toxicity profile:
   - Mavenclad® (cladribine): Generally recommended for inadequate response to alternative drug due to safety profile; used for RRMS and SPMS. Adverse effects include heart failure, hematologic toxicity, hepatic injury, and PML.

(C) Infused Medications

1. For treatment of CIS, RRMS, active SPMS:
   - Briumvi™ (ublituximab-xiiy); monoclonal antibodies. Adverse effects include infusion reactions, infection, and PML.
   - Lemtrada® (alemtuzumab); monoclonal antibodies. FDA recommends generally be reserved if inadequate response to two or more MS therapies. Adverse effects include immune thrombocytopenic purpura, infection, autoimmune disorders, infusion reactions, acquired hemophilia A, acute acalculous cholecystitis, and adult-onset Still's disease.
   - Tysabri® (natalizumab); monoclonal antibodies. Adverse effects include herpes infection hepatotoxicity, PML, and thrombocytopenia.
2. For treatment of SPMS (changed from RRMS to progressive at a variable rate), progressive relapsing MS (gradual increase in disability from onset with relapses), and worsening RRMS (clinical attacks without complete remission - stepwise worsening):
   - Novantrone® (mitoxantrone); antineoplastics: Was used to treat cancer. Adverse effects include dose-related lifetime cardiotoxicity and hematologic toxicity.

3. For treatment of primary progressive multiple sclerosis (PPMS), as well as CIS, RRMS, and ASPMS:
   – Ocrevus® (ocrelizumab); monoclonal antibody. Adverse reactions include immune-mediated colitis, infections, PML, and malignancies.

PML: Rare, progressive white matter brain disease caused by reactivation of a common virus, John Cunningham (JC) virus, which ordinarily causes no symptoms. PML may rarely occur with some medications for MS. Tests are available for JC virus, and can be used before starting treatment and while on treatment to assess risk for PML. Symptoms of PML are similar to MS symptoms and can include motor, visual, speech, and personality.

NONAPPROVED TREATMENTS USED FOR MULTIPLE SCLEROSIS DISEASE MODIFICATION (NATIONAL MULTIPLE SCLEROSIS SOCIETY, 2023)

The side effects need to be weighed when prescribing these medications and patients should be closely monitored. Examples of these medications include:

- Cyclophosphamide (Cytoxan®): Immunosuppressive, may decrease relapse, newer areas of CNS inflammation
- Azathioprine: Immunosuppressive, used in MS over 30 years, with and without other disease-modifying therapies. May decrease relapse rate, decrease new CNS inflammation and disease worsening. Approved in Europe for MS, not in the United States at time of this writing.
- Rituximab (Rituxan®): Monoclonal antibody, IV, may reduce clinical relapses and limit progression and new inflammation.
- Cellcept (mycophenolate mofetil): Immunosuppressive, may reduce number of MS relapses, limit new areas of CNS damage, may slow disease worsening.
- Minocycline: Oral tetracycline, studied in combination with interferon beta-1a and with glatiramer acetate, with the latter showing safety, tolerability, and decreased lesions and T2 burden of disease
- Statins: Approved to lower cholesterol; one statin (simvastatin) found to reduce rate of brain atrophy, while other studies with statins found no benefit as an add-on to beta interferon to reduce activity with RRMS or CIS.

MEDICATION FOR GAIT SPEED

- Dalfampridine (Ampyra®):
  – Broad-spectrum potassium channel blocker
  – Dosing: 10 mg orally twice a day
  – Indicated to improve walking—improves speed in one-third of patients
  – Risk of seizures low

REHABILITATION AND SYMPTOMATIC MANAGEMENT

- **"Rehab improves outcome in MS" (Greenspun et al., 1987).**
  – Weakness and fatigue are seen in the lower extremities >> upper extremities
    ■ A goal of rehabilitation in MS patients is to prevent deconditioning, disuse atrophy, and muscle weakness to maximize functional potential.
- **Exercise improves conditioning.**
  – Aerobic training increases endurance. Light progressive resistive exercises prevent disuse atrophy, allowing for multiple periods of rest.
  – In mild to moderate disability from MS, exercise improves aerobic capacity, strength, mobility, fatigue, quality of life, spasticity, cognition, depression, walking ability, and memory/hippocampal volume.
  – May have neuroprotective effect—Do not exercise to the point of fatigue (submaximal exercise). Evidence-based guidelines for exercise prescription: Mild to moderate disability: 30 minutes of moderate-intensity aerobic activity twice a week and strength training of major muscle groups twice a week.

- **Fatigue worsens with increased temperature, stress, and activity**.
  - Heat worsens the condition due to delayed impulse conduction. Only a slight increase in temperature is needed to result in conduction block.
  - Swimming should be done at a cooler temperature (<84°F).
  - Recommendations: Medications (amantadine, Ritalin®) and avoidance of temperature increases. Provigil shows promise as well. Selective serotonin reuptake inhibitors (SSRIs) may help.

Spasticity Management

- See also Spasticity section in this chapter.
- An increase in spasticity may be an indication of a coexisting condition, including infection, trauma, skin breakdown, and so on.
- **Physical:** Remove noxious stimuli, ROM, positioning, casting and splinting, and cryotherapy. Optimize bowel, bladder, and skin management.
- **Medications: 🕮 Baclofen (oral [PO] or intrathecal) is the drug of choice for central spasticity.** Other medications include tizanidine, dantrolene, diazepam, clonidine, and clonazepam.
- **Local injections:** Motor point, blocks, nerve blocks, and botulinum toxin injections are also used.
- **Surgery:** Rhizotomies, tenotomies, and neurectomies
- **ITB pump:** Baclofen delivered into the intrathecal space via a programmable pump

Incoordination, Ataxia, Tremor, Dysmetria

- Dysmetria is a lack of harmonious action between muscles during a voluntary movement in which the patient is unable to stop motion at a desired point.
- The specific symptoms will depend on the specific location of the plaques in relation to the white matter tracts: Spinocerebellar, cerebrum, and dorsal columns.
- Rehabilitation includes PT/OT for balance training and relaxation techniques. Weighted ankle cuffs and utensils may assist ambulation and ADLs for patients with proprioception abnormalities. Frenkel's exercises are used to treat ataxia.
- Medications used with limited success: Isoniazid, primidone, clonazepam (Klonopin®), Depakote®, Inderal®, and hydroxyzine

Sensory Disturbances and Pain

- PT/OT, therapeutic modalities (e.g., TENS), behavior modification, and trials of various medications may be used to control the sensory dysesthesias that can be associated with MS.
- Medications: Gabapentin (Neurontin®), carbamazepine (Tegretol®), tricyclic antidepressants (TCAs), amitriptyline (Elavil®), nortriptyline (Pamelor®), phenytoin (Dilantin®), capsaicin

Fatigue (Also See "Rehabilitation and Symptomatic Management" Section)

- Overwhelming exhaustion unique from normal fatigue
- Better in morning
- Impacts motor function, cognition, self-care, and so on
- Worse with depression, heat intolerance, vigorous exercise, stress, and spasticity
- Rule out other causes (e.g., anemia, hypothyroidism, medication-induced depression)
- Energy conservation, work simplification, economy of efforts, pacing, and education
- Medications, such as modafinil, amantadine, fluoxetine, and methylphenidate, can be considered to improve fatigue.

Visual Impairment/Ocular Findings

OPTIC NEURITIS

- Acute (hours to days) inflammatory demyelination of the optic nerve. Seen in one-fourth of all MS patients.

- **Clinical presentation:** Blurred vision, or complete or partial loss of vision, may be found in one eye. Usually unilateral eye pain is present, which is worse with eye movement. Vision loss may be preceded by pain around the eye. A central scotoma is a common visual field defect. Residual decreased acuity, photophobia, or pain possible.
- **Treatment:** IV methylprednisolone in selected cases. Long-term vision is not affected by treatment.

INTERNUCLEAR OPHTHALMOPLEGIA

- Demyelinating lesion of the medial longitudinal fasciculus (MLF)
- **Clinical presentation:** Paresis of the medial rectus muscle, resulting in an inability to adduct the eye; this is seen with voluntary lateral gaze. This lesion is usually accompanied by nystagmus.
- Convergence, which does not run in the path of the MLF, involves a different pathway and is intact. Convergence runs from the retina to the midbrain nuclei via the pathway of the optic nerve, tract, chiasm, and lateral geniculate terminating in the bilateral cranial nerve (CN) III nuclei.
- Nystagmus, diplopia, blurred and decreased vision
- **Treatment:** Patching and prisms

Bladder Dysfunction (Table 12–15)

- Bladder dysfunction is common in MS patients and occurs due to the development of plaque lesions affecting the nerve signals in the CNS that control bladder function. Also see Spinal Cord Injury section on bladder management in Chapter 7, Spinal Cord Injuries, for an in-depth discussion.

Other Abnormalities

- **Bowel dysfunction:** Constipation is commonly seen. An early bowel program is recommended.
- **Dysphagia:** Impairment of CNs (V, VII, IX, XII) may lead to swallowing abnormalities. Delayed swallowing, dysphagia, or pooling may be seen. A swallowing evaluation by speech therapy is essential in preventing complications such as aspiration. Treatment includes exercise, positioning, and a change in food consistency.
- **Speech disorders:** Dysarthria, dysphonia
- Sexual dysfunction
- **Cognitive/affective impairments:** Process speed, memory, executive function
  - General intelligence in MS changes gradually over time, and verbal skills are affected less than performance skills.

TABLE 12–15 Neurogenic Bladder Types in MS

	FAILURE TO STORE (MOST COMMON)	FAILURE TO EMPTY	COMBINATION BLADDER
Disorder	• Hyperactive bladder with a small capacity and weak sphincter (adrenergic)	• Big boggy bladder • Closed sphincter	• Known as DSD
Problem	• Incontinence • Urinary frequency, dribbling of urine	• Failure to void • Overflow incontinence	• Bladder contracts • Sphincter closes • Backflow of urine to the kidney
Treatment	*Detrusor muscle relaxant examples* • Ditropan® • Tolterodine (Detrol®) • Mirabegron (Myrbetriq®)	• Self-intermittent • Catheterization placement *Detrusor muscle contractor* • Urecholine: External sphincter relaxant *Alpha–antagonist* • Minipress® • Flomax®	• Intermittent catheterization • Anticholinergics for storing between catheterization • Alpha-antagonists used by some • Urethral stent or sphincterotomy

DSD, detrusor sphincter dyssynergia; MS, multiple sclerosis.

  - Interferon beta-1a and 1b, natalizumab, and fingolimod may slow progression.
  - 🕮 The IQ of MS patients is usually intact, especially early on. Processing speed is often delayed (Peterson & Kokmen, 1989).
  - Decreased short-term memory, decreased reasoning, and slow processing—Euphoria may be caused by frontal lobe lesions or corticosteroid use—Cortical atrophy correlates with cognitive impairment
- Up to 70% have neuropsychiatric abnormalities
  - Depression is a common finding and should be addressed by rehab psychology; impacts memory, attention, and concentration.
  - Suicide rate is 7.5 times higher than normal population.
- Pseudobulbar affect: Uncontrollable laughter or crying incongruent with situation

OUTCOMES IN MULTIPLE SCLEROSIS

- On average, longevity is approximately 5 to 10 years shorter in people with MS.
- Seldom fatal
- One-third require ambulatory assistance within 10 years of diagnosis; two-thirds do not.
- 59% of patients with PPMS require unilateral walking assistance within 10 years (Kerbrat et al., 2015).

Minimal Record of Disability

- This consists of different types of rating scales that profile the main dysfunctions of MS.
- The most common scale used is the Kurtzke Expanded Disability Status Scale (EDSS).
  - A 10-level rating scale used in MS examining eight different neurologic systems.
  - Rating scale:
    0 = normal
    4 = severe disability, but still ambulatory without aid
    8 = bedbound
    10 = death
  - Areas tested: Pyramidal, cerebellar, brainstem, sensory, bowel and bladder, vision, mental status, and general
- Other outcome scales used in MS include:
  - Kurtzke Functional Systems (FS)
  - Incapacity Status Scale (ISS)
  - Environmental Status Scale (ESS)
- Minimal record of disability (MRD) = EDSS, ISS, and ESS
- FIM: Assesses disability and the need for assistance. Does not assess vision (Kurtzke, 1983; see **Figure 8–4**).

MUSCULOSKELETAL ULTRASOUND

BACKGROUND

- The use of US in PM&R has become commonplace. Its popularity has grown due to recent advances in technology, which have produced higher resolution images in more portable and affordable systems.
- US should serve as an extension of the clinical exam as it offers value in the diagnosis and management of musculoskeletal pathology.

- Advantages of US compared with MRI:
  - Improved soft tissue differentiation
  - Dynamic, real-time evaluation: Allows direct visualization of ligaments/tendons and their integrity while performing provocative tests (e.g., US imaging of the medial collateral ligament during valgus stress testing of the knee)
  - Sonopalpation: Visualization of tissue response to palpation with the US transducer
  - Ability to perform side-to-side comparisons
  - Lower cost
- Disadvantages:
  - Imaging results and interpretation are user-dependent, and significant expertise is necessary
  - Poor visualization of intra-articular and subcortical bony pathology due to acoustic reflection off bone (e.g., sound waves do not penetrate well through cortical bone)
    - MRI is far superior in evaluating intra-articular pathologies (anterior collateral ligament, posterior collateral ligament, meniscal, or labral tears) as well as stress fractures.
  - Study depth limitations (image quality can be problematic with deeper structures and/or in patients with larger body habitus)

HOW ULTRASOUND WORKS

- The US machine comprises two main parts: The computer and the transducer.
- The transducer contains piezoelectric crystals that are responsible for both the generation of sound waves (**reverse piezoelectric effect**) and detection of the returning echoes to form images (**piezoelectric effect**).
  - Electric signal ↔ Transducer crystal ↔ Sound wave
- The choice of transducer is usually based on the size and depth of the structure being evaluated. The most common frequency ranges are between 5 and 12 megahertz (MHz).
- Commonly used transducers:
  - **Linear array transducer**: High frequency (5–12 MHz), higher resolution images, poorer depth penetrance. Ideal for superficial structures (e.g., hand/wrist/elbow, shoulder, knees).
  - **Curvilinear array transducer**: Low frequency (3–5 MHz), lower resolution images, greater depth penetration. Ideal for deeper structures (e.g., hip joint, in patients with large body habitus).
  - **Compact linear ("Hockey-stick") transducer**: (≥10 MHz), very high frequency, very high resolution, superficial penetrance. Ideal to visualize superficial structures (e.g., finger, median nerve).
- Transducers provide thin, 2D images, of a 3D structure. For this reason, a sonographer must ensure complete structural evaluation to avoid missing pathology.

BASIC ULTRASOUND TERMINOLOGY

- **Wavelength** is the distance between consecutive cycles of sound waves.
- **Frequency** is the number of complete cycles produced per unit of time.
  - Commonly measures in hertz (Hz) = One wave cycle per second
  - 1 MHz = 1,000,000 Hz
- When pulses of the sound wave pass through tissues, distinctive images are formed that are dependent on the amount of energy absorbed or reflected.
- Important factors include tissue density, acoustic impedance, and propagation speed of the sound wave. **Reflection** of the sound waves occurs at the boundary between two structures.
- **Resolution:** The ability to differentiate between structures that are closely related
  - **Higher frequencies** generate higher image resolution and detail but are unable to penetrate as deeply into the body.
  - **Lower frequencies** generate lower resolution images but are able to penetrate deeper into the body.

- **US views:**
  - **Longitudinal view:** Transducer is oriented lengthwise (parallel) to the long axis of the structure or body part being examined.
  - **Transverse view:** Transducer is oriented across (perpendicular to) the long axis of the structure or body part being examined.

ULTRASONOGRAPHIC CHARACTERISTICS OF TISSUES

- Understanding ultrasonographic characteristics of tissue is essential for both tissue differentiation and identification of pathologic states **(Figures 12–14 to 12–17)**.
- Important ultrasonographic characteristics include echogenicity, anisotropy, echotexture, compressibility, and blood flow.
  - 🕮 **Echogenicity:** This refers to the ability of a structure to reflect sound waves and produce echoes. It is graded and described on the gray scale.
    - **Hyperechoic:** Structure highly reflects sound and is brighter than the surrounding tissues (white).
    - **Isoechoic:** Structure has the same echogenicity as the surrounding tissues (light gray).
    - **Hypoechoic:** Structure minimally reflects sound and is darker than surrounding tissues (dark gray).
    - **Anechoic:** No sound reflection (black)
  - 🕮 **Anisotropy (Figure 12–14):** A directionally dependent property of certain tissues, in which a single tissue will have variable appearance based on the angle/direction of the sound waves**.**
    - For example, there will be loss of the normal hyperechoic appearance of tendon when the transducer beam is not perpendicular to the structure. Slight angulation causes the sound wave to refract away from the transducer ("drop off") and appear darker.

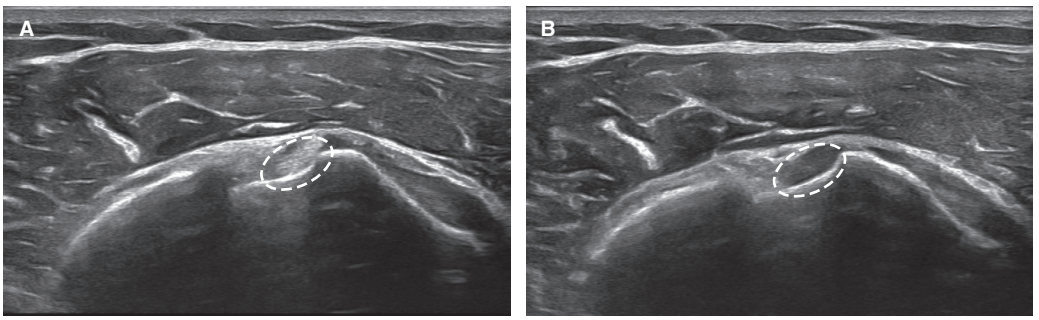


FIGURE 12–14 (A) Biceps tendon in bicipital grove (inside dashed lines). (B) Anisotropy of biceps tendon (inside dashed lines).

  - **Echotexture:** This refers to the internal echo pattern of a tissue. A single tissue type can have variable echotexture based on probe positioning (this is described in the text that follows).
  - **Compressibility:** Veins are more compressible than arteries. Arteries will continue to be pulsatile despite compression.
  - **Blood flow:** The presence of blood flow in an avascular structure can be suggestive of a pathologic state (e.g., neovascularization in tendinopathy).

🕮 OVERVIEW OF ULTRASOUND APPEARANCE BY TISSUE

- **Bone:**
  - Bone is highly reflective of sound and will appear hyperechoic (bright) with acoustic shadowing (lack of signal) deep to it.
  - Calcium deposits (e.g., calcific tendinitis, HO) in other areas of the body will have the same characteristic image findings.

- **Soft tissue:**
  - **Tendons:** Highly organized linear strands that will appear hyperechoic (Figure 12–15)
    - Transverse view: Echogenic dots; "broom-end" appearance
    - Longitudinal view: Fibrillar pattern
    - Tendon is most susceptible to anisotropy, and caution must be used as anisotropic artifact may be misinterpreted as a pathologic state. A loss of fibrillar pattern is characteristic of tendinosis and an anechoic defect may suggest a tendon tear.

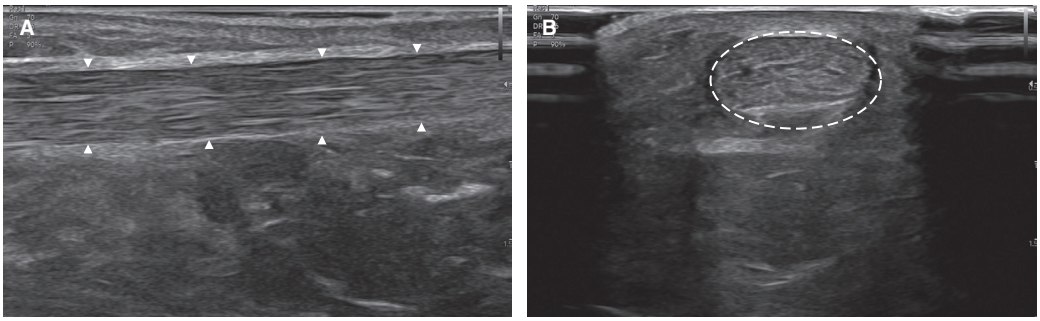


FIGURE 12–15 Achilles tendon in (A) longitudinal view (arrows) and (B) transverse view (inside dashed line).

  - **Nerves:** Appear as tubular structures with mixed echogenicity: Hyperechoic fascicles within a hypoechoic background (Figure 12–16)
    - Transverse view: Speckled or "honeycomb" appearance; also resembles a cable in cross section
    - Longitudinal view: Fascicular pattern

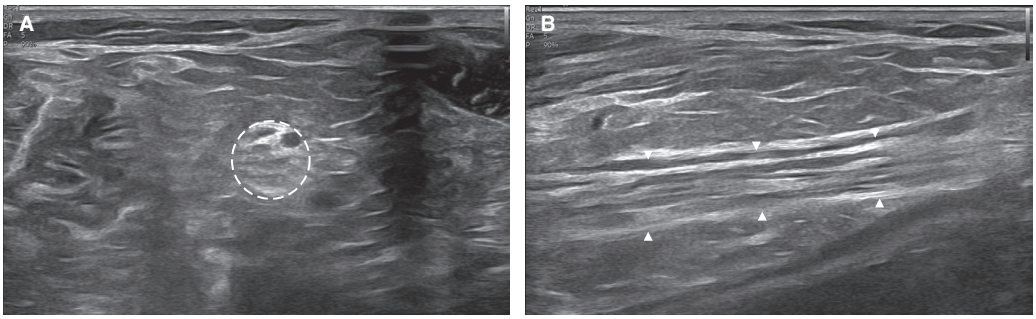


FIGURE 12–16 Sciatic nerve in transverse (A) view (inside dashed line) and longitudinal (B) view (arrows).

  - **Ligaments:** Hyperechoic with a more compact fibrillar pattern than tendons
  - **Skin:** Slightly hyperechoic
  - **Muscle:** Mixed echogenicity: Hypoechoic areas surrounded by organized, hyperechoic linear septae and surrounding epimysium (Figure 12–17)

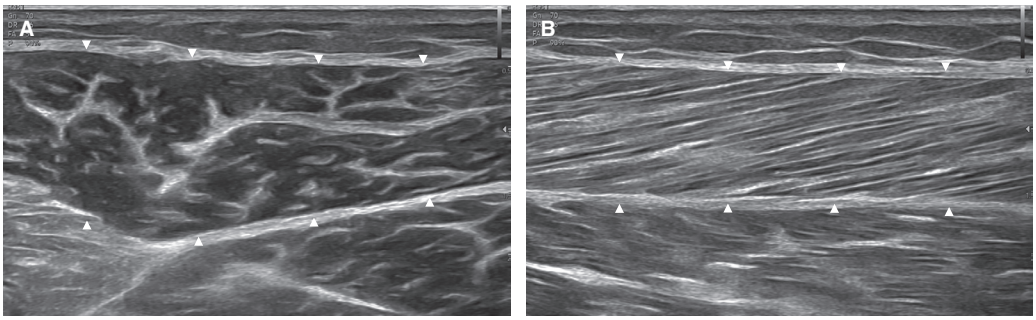


FIGURE 12–17 Gastrocnemius muscle (A) in transverse view and (B) in longitudinal view (arrows).

  - Longitudinal view: Pennate pattern
  - Transverse view: "Starry night" appearance
- **Fat:** Usually hypoechoic interspersed between hyperechoic, irregular, septated lines
- **Cartilage:** Hypoechoic to anechoic
- **Fluid:** Anechoic

ULTRASOUND IMAGING ARTIFACTS

- US artifacts are routinely encountered during the scanning process and can cause misinterpretation of an image.
- **Posterior acoustic shadowing**: Decreased or loss of sound wave signal due to strong reflection (bone, large calcifications) or strong attenuation (solid tissue, dense/malignant masses)
- 🕮 **Anisotropy:** See above section "Ultrasonographic Characteristics of Tissues."
- **Posterior acoustic enhancement (enhanced through transmission)**: Refers to increased echogenicity of tissue that is deep to fluid due to sound waves traveling well through fluid
- **Reverberation**: When the surface of an object is smooth and flat, the sound beam reflects back and forth between the surface and transducer. The result is a series of linear echoes that extend deep into the structure. Commonly seen with metal surfaces or implants.

ULTRASOUND-GUIDED PROCEDURES

- US image guidance provides improved procedural safety by allowing direct and real-time visualization of structures.
- Multiple studies document improved accuracy of needle placement (Jackson et al., 2002; Naredo et al., 2004).
- Common US-guided procedures:
  - Diagnostic and/or therapeutic joint, bursa, and tendon/tendon sheath injections
  - Arthrocentesis and bursa aspiration
  - Peripheral nerve blocks and chemodenervation
  - Percutaneous tenotomy/fasciotomy

NEEDLE INSERTION METHODS

- **Indirect method:** US is used to localize ("mark") the needle insertion site and confirm the target location. This technique does not use real-time US guidance during needle placement and advancement.
- **Direct method:** The needle is visualized and advanced in real time under US. This allows the practitioner to direct the needle to the target area.
- Needle visualization depends on the alignment of the needle with the US beam. Newer US machines offer needle enhancing software that improves needle visualization.
- **Needle insertion techniques:**
  - **Long axis/in-plane approach:** The needle is placed in *parallel* (colinear) with the transducer. This approach allows the entire needle to be visualized throughout the procedure.
  - **Short axis/out-of-plane approach:** The needle is inserted *perpendicular* to the transducer. The length of the needle cannot be directly visualized. When visualized, the needle can appear as a white dot. Indirect target area confirmation can be noted with fluid expansion into the area during the injection (e.g., enlarged bursa with injectate administration rather than in muscle).

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EPILOGUE

I hope you enjoyed this textbook and that it has helped crystallize the facts needed to pass boards and practice the field of physical medicine and rehabilitation. It was not meant to be the final text but a complete factual outline to enable both the seasoned and the neophyte to gain a vast amount of knowledge and background pathophysiology of diseases that cause disabling conditions. It will serve as an excellent quick reference to you for years to come. One must always remember, "We see what we are looking for, we look for what we know, and what we do not know, we never see." Learning is a lifelong experience. Enjoy the experience and continue looking for the unseen.

Thomas E. Strax, MD

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